DOCUMENT RESUME

ED 437 763 EC 307 611

AUTHOR	Houghton, Joan; Coonts, Teresa; Jordan, Beth; Schafer, Jacqueline, Ed.
TITLE	Screening for Usher Syndrome: A Hands-On Guide for School Nurses.
INSTITUTION	Kansas State Board of Education, Topeka.
SPONS AGENCY	Special Education Programs (ED/OSERS), Washington, DC.
PUB DATE	1999-00-00
NOTE	61p.; In collaboration with Mae Claxton, Basil Kessler, and Joyce Markendorf. Edited by Jacqueline Schafer. For more information on students with dual sensory impairments, see ED 393 275, EC 307 610 and EC 307 612. Includes Usher Screening Forms in Spanish.
CONTRACT	H025A50015
AVAILABLE FROM	Kansas Dual Sensory Impairments Project, Kansas State Department of Education, 120 SE 10th Ave., Topeka, KS 66612. Tel: 785-296-0917 (Voice/TTY); Fax: 785-296-1413; e-mail: jhoughton@ksbe.state.ks.us.
PUB TYPE	Guides - Non-Classroom (055)
EDRS PRICE	MF01/PC03 Plus Postage.
DESCRIPTORS	Case Studies; *Disability Identification; Elementary Secondary Education; Evaluation Methods; *Hearing Impairments; Information Sources; Neurological Impairments; *School Nurses; *Screening Tests; Student Characteristics; Student Evaluation; Symptoms (Individual Disorders); *Visual Impairments
IDENTIFIERS	*Related Services; *Usher Syndrome

ABSTRACT

This manual was written specifically to help school nurses conduct screenings for Usher syndrome, a genetic condition that involves deafness or hearing loss and the progressive loss of vision. It provides information on the step-by-step process of how to conduct a screening, the actual forms needed for a screening, and resources for referring students for further evaluations and counseling. Sections of the manual address: (1) symptoms of Usher syndrome; (2) heredity of the disorder; (3) how a screening is conducted; (4) conducting a paper screening; (5) station screening, the role of the screener, and the role of the interpreter; (6) evaluating the results; (7) when to recheck a student; and (8) when to refer a student for an opthalmological evaluation. Written and visual resources are provided, along with national, state, and regional organizations relating to Usher syndrome. Appendices include supplemental materials that address retinitis pigmentosa, characteristics of Usher syndrome and sensorineural hearing loss, electroretinogram evaluations, an Usher screening inservice agenda, and how to explain Usher syndrome to kids. A case study of a student with Usher syndrome is provided, and blank forms include questionnaires, the Cone Adaptation Test, the Field Screening Test and the Balance Test. (Contains 32 references.) (CR)

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SCREENING FOR USHER SYNDROME

A hands-on guide for school nurses

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Screening for Usher Syndrome
A hands-on guide for school nurses

By:
Joan Houghton, MS Ed
Kansas State Department of Education
Teresa Coonts, MS Ed
National Technical Assistance Consortium
Beth Jordan, M Ed
Helen Keller National Center

With:
Mae Claxton, RN
Kansas State School for the Blind
Basil Kessler, M Ed
Kansas State Department of Education
Joyce Markendorf, RN, MSN
Kansas Department of Health & Environment

Edited by:
Jacqueline Schafer, MS

Kansas Services for Children and Youth with Dual Sensory Impairments, Project #H025A50015, is funded by the Office of Special Education Programs, U.S. Department of Education. Information obtained within this publication does not necessarily reflect the views of the U.S. Department of Education.

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Acknowledgments

This effort would not be possible without the collaboration and sharing of many individuals. Their hard work, time commitment, and willingness to complete various tasks are reflected in this document.

The Kansas Usher Screening Committee wants to acknowledge foremost the students from Kansas State School for the Deaf and Wichita Public Schools who participated in the Usher Screening Pilot. Other individuals who deserve recognition for their contributions are Dr. Sandra Davenport, Project Coordinator; Dr. Sara Gaar, Alaska Dual Sensory Impairments Project; Earlene Dykes, Arizona Deaf-Blind Project; Barb Sims, Illinois Center on Deafness; Mariel Zeller, North Dakota Department of Education; Dr. Brent Askvig, North Dakota Deaf-Blind Project; Lisa Jacobs, New England Regional Center; Marta Vinton, Usher syndrome Support Group; Sandra Andrews, Envison; Bill Daugherty and Virgene Tyrell, Kansas State School for the Blind; Gerald Johnson, Carmen Norris, and Mary Montgomery, Kansas State School for the Deaf; Dr. Trisha Self, her graduate students, and Kara Kissick, Wichita State University-Department of Speech Pathology and Audiology; Terry Baccus and Molly Potroff, Wichita Public Schools; Basil Kessler, Transition Statewide Systems Change Project-Kansas State Department of Education; and Marnie Campbell, Kansas State Department of Education.

To all of you—many, many thanks.

The members of the Kansas Usher Screening Committee

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Preface

This manual was written specifically to help school nurses conduct screenings for Usher syndrome, a genetic condition that involves deafness or hearing loss and the progressive loss of vision. As such, it provides the step-by-step process of how to conduct a screening, the actual forms needed for a screening, and resources for referring students for further evaluations and counseling.

Please be aware that members of the Kansas Usher Screening Committee are available to help conduct Usher syndrome screenings. To request on-site assistance, nurses should contact the Director of the Kansas Dual Sensory Impairments Project, Kansas State Department of Education, Student Support Services Division, 120 SE 10th Avenue, Topeka, Kansas 66612; (785) 296-0917 (V/TTY).

The Kansas Usher Screening Committee was formed in 1996 in response to a growing national awareness of the importance of screening for Usher syndrome. The committee began meeting in 1996 to learn how to conduct educational screenings for students who may have Usher syndrome. Committee members met for a year under the guidance of Dr. Sandra Davenport, a leading national expert who pioneered educational screening efforts for Usher syndrome. Two pilot site screenings were conducted: one at Kansas State School for the Deaf and one in Wichita Public Schools. A total of 79 students participated in the screenings. Of this total, 37 students passed both paper and site screenings, seven students were referred for an ERG, 25 students were targeted for rechecks within one year, and 10 students were targeted for vision evaluations other than an ERG. One of the two students who were referred for an ERG was diagnosed with Usher syndrome.

Preface

A collaborative effort between the Kansas Department of Health and Environment and the Kansas State Department of Education, Student Support Services, was initiated to include testing for Usher syndrome with the vision screening certificate training offered every year at various locations throughout the state. The training combined with the development of resource packages on Usher syndrome and this procedural manual should help increase the awareness and implementation of screening programs. Through this collaboration from members of the Kansas Usher Screening Committee, students who have this syndrome and their families will receive appropriate educational services and supports.

Section I:

Introduction

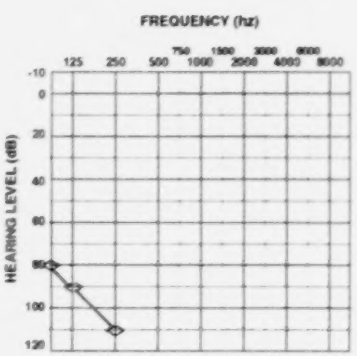
What Is Usher Syndrome?

Usher syndrome is a genetic condition that involves deafness or hearing loss and the progressive loss of vision due to Retinitis Pigmentosa (RP)[1]. In addition, depending on the type of Usher syndrome acquired, it may affect a person's sense of balance.

There are three types of Usher syndrome. Different characteristics of vision, hearing, and balance are associated with each type. (For a list of the different characteristics associated with each type of Usher syndrome and a description of the different types of hearing loss, see page 27.)

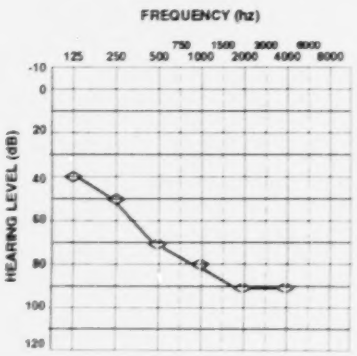
A person with Usher syndrome Type I typically is born with a profound hearing loss in both ears, which is characterized by a corner audiogram. (See Diagram 1 for an illustration of a corner audiogram). A visual field loss usually is detected in early elementary years and progresses to total blindness as an adult. Finally, a person's sense of balance is affected.

Diagram 1: Corner Audiogram



A person with Usher syndrome Type II typically is born with a moderate to profound hearing loss in both ears, which is characterized by a sloping audiogram. (See Diagram 2 for an illustration of a sloping audiogram.) Vision loss usually occurs during the mid to late teenage years and does not usually progress to total blindness. The person's balance is not affected.

Diagram 2: Sloping Audiogram



[1]For more information on Retinitis Pigmentosa, see "What is Retinitis Pigmentosa?," page 26.

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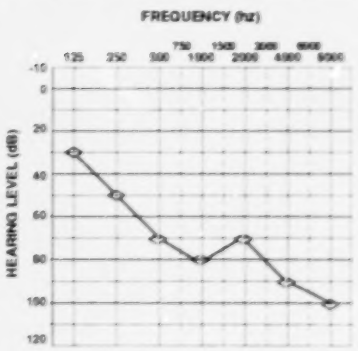
Section I:
Introduction

A person with Usher syndrome Type III (the most rare) typically is born with normal hearing or a mild to moderate hearing loss in both ears; however, the hearing loss may drop drastically within a five- to ten-year period. This loss is characterized by an audiogram with a slope and a ski bump. (See Diagram 3 for an illustration of this type of audiogram.) Vision loss progresses the same as Usher syndrome Type II; however, legal blindness may occur from 20 to 40 years of age. Some individuals may experience balance problems.

Diagram 3: Ski Bump Audiogram



How Does a Person Inherit Usher Syndrome?

Usher syndrome is an "autosomal recessive condition." "Autosomal" means that the genes are not sex linked. Males and females are affected equally. "Recessive" means that the Usher gene is inherited from each parent. Most parents do not know they are a carrier of Usher syndrome until their child is diagnosed. The possible combinations that will result in Usher syndrome are:

- If both parents are carriers, the chances are one in four children will have Usher syndrome.
- If only one parent has Usher syndrome and the other is not a carrier, no children will be affected; however, all will be carriers.
- If one parent has Usher syndrome and the other parent is a carrier, two of four children (50%) will have Usher syndrome.
- If both parents have Usher syndrome, all of the children will have Usher syndrome.

Why Is Screening Important?

Screening for Usher syndrome is important because students with Usher syndrome usually require specific accommodations and

Section I:
Introduction

modifications in order to function effectively and safely. They also have behaviors that are often misinterpreted as clumsy, slow, and uncooperative.

Thus, screening may identify students who need to learn how to safely function within different environments, students who need additional related services in school (e.g., career planning, genetic counseling, and social or emotional counseling), and students who need modifications and adaptations during instruction. Once Usher syndrome is diagnosed, students can better prepare for the changes facing them as the condition progresses.

Caution: Screening can only *detect* early signs of Usher syndrome. For an accurate diagnosis, an ophthamological evaluation of an electroretinogram (ERG) and extensive visual field testing are required. (For more information on ERGs, see "What is an Electro-retinogram Evaluation?" page 28.) Additionally, genetic counseling is recommended to confirm the student has Usher syndrome.

How is the Screening Conducted?

Basically, there are two parts to a screening: the paper screening and the station screening. The purpose of the paper screening is twofold: a) to target candidates for the station screening, and b) to eliminate students from participation in further screenings. The paper screening identifies family history that may indicate the potential for Usher syndrome and behavior characteristics that may indicate a visual field loss, blind spots, or acuity loss. (Refer to pages 5-8 to learn how to conduct a paper screening.)

The station screening consists of three stations: visual field, cone adaptation, and balance. The visual field and cone adaptation stations are used to identify students who demonstrate visual behaviors that are characteristic of RP, rod deterioration, or cone deterioration. The balance station is used to identify students who demonstrate balance difficulties with the lights on or the lights off. (Refer to pages 9-15 to learn how to conduct a station screening.)

Section I:
Introduction

Help! Where Do I Begin?

Before you begin a screening, familiarize yourself with the different types of Usher syndrome, the inheritance patterns, and why screenings are important (pp. 1-3). Next, do the following:

1. Read Sections II, III, and IV of this manual.
2. Copy the forms needed for the paper screening. These are listed on page 5.
3. After the paper screening has been conducted, determine if a station screening is required. (See pp. 5-8 for making this determination.)
4. Obtain a *Cone Adaptation Test* if you do not have one. (See p. 25.)
5. Copy the forms necessary for the station screening. (See p. 8.)
6. Determine if you need help conducting the station screening. If you do, contact the Director of the Kansas Dual Sensory Impairments Project, Kansas State Department of Education, 120 SE 10th Avenue, Topeka, Kansas 66612; (785) 296-0917 (V/TTY). Remember: Members of the Usher Screening Committee are available to help.
7. Evaluate the results from the paper screening and the station screening. Determine referral status of the student by following the instructions in Section IV. If you are not sure about the results, read the case study in Appendix B.
8. Review resources in Section VII. Refer school administrators, teachers, and family members to experienced evaluation personnel located in this section.
9. Once the student has been diagnosed, give school administrators, teachers, family members and, if appropriate, the student (age 18 and his or her own guardian) *A Collection of Resources on Usher Syndrome Packet* that contains useful information on Usher syndrome. (Refer to Section VII to obtain a packet.)
10. Contact the Director of Kansas Dual Sensory Impairments Project for processing certification on the Kansas State Deaf-Blind Census.

Section II:

The Paper Screening

Prior to the Paper Screening

A. Identify students who have the characteristic hearing loss as demonstrated by audiogram (see Diagrams 1-3, pp. 1-2).

B. Send a letter from the school to parents/guardians of these students. Include the following materials from Appendix C:

- *Kansas Usher Screening Project Family Questionnaire*
- *Kansas Usher Screening Project Behavior Questionnaire*
- Your district's release of information form
- Self-addressed stamped envelope, if possible.

Timeline:
Allow two weeks from the date of the parent/guardian letter for families to return materials. If materials are not returned within this time, contact families by telephone to encourage their involvement in the screening process.

Upon Receipt of the Family Information

A. Obtain the following completed information:

- *Kansas Usher Screening Project Behavior Questionnaire* (from the student's teachers and dorm staff, if appropriate)
- *Kansas Usher Screening Project Student Questionnaire*
  Note: the Student Questionnaire should be completed for students from 3rd grade through 12th grade. The student's teacher should interview the student to complete the form.

Timeline:
Allow one month to obtain the above questionnaires.

After You Have Received All Materials

A. Review all materials and determine if the student should receive a station screening. Students who meet at least HALF of the criteria on each of the following items should receive a station screening.

Section II:

The Paper Screening

Item #1: Family History Questionnaire

- The student has not been diagnosed with another syndrome (e.g., Goldenhar or Rubella).
- The student did not have multiple organs effected at the time of birth or as a result of a degenerative condition.
- The student's family history indicates no multiple generations of deafness or hard of hearing, or neither biological parent is deaf or hard of hearing.
- There is no incidence of mental retardation.
- The student walked earlier than 18-24 months.
- The student was born deaf or hard of hearing (bilateral sensorineural loss). Or, the student was born with normal hearing and it dropped significantly within a 5-10 year period and that drop was not the result of infection, disease, or trauma.
- The student demonstrates visual behaviors that indicate difficulty seeing at night, in dim light, or a person signing from the side of his or her body.
- The student has balance difficulties when walking on uneven surfaces, bumps into objects like tables and chairs, or knocks things over when reaching for them.
- The student has no noticeable physical characteristics (e.g., white patch of hair) or other health issues that could possibly indicate another syndrome (e.g., Refsum) that could result in deaf-blindness.

Note: If both parents are hearing or one is hearing and one is deaf AND there are one or two relatives from past generations who were born deaf or hard of hearing and lost their sight in their midteens to early 30s, the student should receive a station screening. If both biological parents have Usher syndrome, the student definitely needs to receive a station screening.

Item #2: Audiogram

- The student's audiogram shows no gradual changes over time.
- The shape of the student's audiogram is sloped or dropped.
- The slope of the loss depicted on the student's audiogram does

Section II:

The Paper Screening

not rise in the higher frequencies.
- The audiogram only shows one ear with a unilateral, sensorineural loss.

Note: A corner audiogram with losses in the severe to profound range and lower frequencies, a sloping audiogram, or a sloping audiogram with a ski bump would indicate a need for a station screening.

Item #3: Student Behavior Questionnaire

- The student bumps into objects, misses curb cuts, or misses the top of stairways.
- The student turns his or her head while reading or uses a finger to mark a spot on the page.
- The student does not respond to signing or waving to the side of his or her body.
- The student is not able to see cars approaching from the side of his or her body at an intersection.
- The student is not able to function in dim light or is night blind.
- The student has problems adjusting to environments ranging from brightly lit to dimly lit.
- The student does not fully participate in groups, does not like new situations, and is the last to enter a room.
- The student is unable to ride a bike.
- The student is unable to function in low contrast situations.

Note: If more than one person marks the same behaviors on each section of the form, this may indicate a visual field loss or balance difficulties. The student should be referred to a station screening.

Item # 4: Student Interview Questionnaire (if applicable)

- The student reports tripping over objects on the floor.
- The student reports being unable to adjust to light within a couple of seconds when entering a darkened area (e.g., movie theater).

- The student reports being unable to see the stars at night.
- The student reports being unable to see moving objects from the side of his or her body (e.g., friends waving, cars moving, or individuals signing).
- The student reports difficulty riding a bicycle.
- The student reports bumping into things or knocking over objects placed on a table.

Note: A large number of matching responses (e.g., half or more) on the *Student Interview Questionnaire* and the *Student Behavior Questionnaire* may indicate visual field losses or balance difficulty. The student should receive a station screening.

B. Develop pocket folder for each student to be screened with the following information in each folder:

Completed Forms:

- Student audiograms (current and previous)
- *Family Questionnaire*
- *Behavior Questionnaires* (teacher(s), parent, and dorm teacher if student lives in residential setting)
- *Release of Information*

Blank Forms:

- *Cone Adaptation Form*
- *Balance Screening Form*
- *Field Screening Form*
- *Kansas Usher and Vision Screening Data Summary Form*

Section III:

The Station Screening

Preparation

A. Set date(s) for screening in collaboration with school schedule so there are no conflicts with field trips, enrichment experiences, assemblies, or other school activities.

B. Plan for the screening day. This includes:
   1. Gather the following materials:
      - *Cone Adaptation Test*
      - Light meter
      - 2 timers or stop watches
      - Students' pocket folders
   2. Arrange for interpreters to match communication needs of the students (e.g., ASL, SEE, or CASE).
   3. Identify rooms for each station (i.e., cone, field, and balance) and determine if appropriate:
      a. Cone: Can the room be made dark enough for testing?
      b. Field: It should be a small room with good lighting and three chairs (evaluator, interpreter, and student).
      c. Balance: It should be a room large enough for two evaluators, two interpreters, and two students. Each student should be able to extend both arms out to the side of his or her body.
   4. Recruit people to assist with the screening (Contact the director of the Kansas Dual Sensory Impairment Project for help in identifying possible screeners).
   5. Develop a schedule for screening (e.g., screen elementary, middle school, high school students separately; groups should consist of no more than four students at one time).
   6. Inform teachers of time, day, and location of the screening in order to release students from class.
   7. Hold a planning meeting with all screeners to run through the screening process. (See example of Usher Screening Inservice Agenda, p. 29.)
      a. Identify who will work at each station.
      b. Identify someone to assign students to stations and give them folders when they first enter the screening area.
      c. Explain how to complete the forms.
      d. Have screeners practice the process on each other.

Section III:
The Station Screening

e. Complete any of the tasks that require a great deal of time (e.g., review completed questionnaires to determine which students may have difficulties during the station screening).

The Role of the Screener

Screeners who are identified to assist in the screening process must have adequate information about Usher syndrome, how it occurs, behavioral characteristics, and psycholsocial and educational ramifications. To avoid burdening school nursing staff, screeners can be recruited from within the district or from the Kansas Usher Screening Committee. Examples of possible district staff include consulting or on-staff speech therapists, audiologists, teachers of vision or hearing, and medical doctors or nurses in the neighboring community. For assistance from the Kansas Usher Screening Committee, contact the director of the Dual Sensory Impairments Project at the State Department of Education (see p. 24 for contact information).

The total number of screeners needed will depend upon the number of stations and number of students needing the station screening. If only a handful of students require the station screening, only two or three screeners will be needed. However, if more than five students will receive the station screening, at least one screener should be identified for each station.

To insure consistency and accuracy of test results, it is essential that screeners meet in advance of the station screening to learn about Usher syndrome, understand the purpose of screening, and develop clear expectations for the screening proces by practicing the various tests on each other. A sample in-service agenda is included on page 29 to facilitate preparation for the station screening day.

Section III:
The Station Screening

The Role of the Interpreter

In order for the screening process to be valid, interpreters must be provided for students who rely on a visual, tactual, or oral interpreter in the classroom. In addition, interpreters must match the communication needs of the student (e.g., students utilizing American Sign Language (ASL) paired with ASL interpreters; students utilizing a Signed English system paired with interpreters trained in the student's sign language system). Likewise, if a student relies on amplification, an assistive listening device (ALD), glasses or other visual aids, these must be used during the screening process.

The total number of interpreters needed for the screening will depend upon the number of screeners at each station. The number of students being screened and the room layout will determine the number of screeners and interpreters. The approximate number of interpreters needed for each activity is:

Activity	**Number of Interpreters**
Welcome/Explanation	1
Cone Adaptation	1-2
Balance	2
Field Screening	1
TOTAL	5-6

Interpreters should be assigned to one station and remain there during the duration of the screening process. This will minimize confusion and ensure consistency. Interpreters should arrive 15 minutes before the screening process begins. The screener for each station will explain the testing process to the assigned interpreter. Breaks should be provided every 30 to 60 minutes so interpreters can refresh themselves. For tactile interpreting, breaks should be provided every 15 to 30 minutes.

Section III:
The Station Screening

The Screening

A. Welcome students and explain procedures

People and Materials Needed:
- Greeter (greets students, gives them their folder, and directs them to stations)
- Explainer (someone familiar with the screening process)
- Mock student (another staff person to role-play)
- Interpreter (using communication skill of the students)
- *Cone Adaptation Test*

Room Setup:
Select a room large enough to accommodate an explainer, an interpreter, a mock student, and up to four students. During this activity, students may choose to stand or be seated. However, students must be able to clearly see the role play and the interpreter.

Procedures:
The greeter welcomes the students (there should be no more than four students in each group) and hands each of them their student folder. The greeter then directs all of the students into the room for the explanation and demonstration.

The explainer briefly describes Usher syndrome and explains the screening process to the students. (See "How to Explain Usher Syndrome to Kids," pp. 30-31.) The explainer and mock student then demonstrate each screening activity (i.e., *Cone Adaptation Test*, the *Balance Test,* and the *Field Screening*). Care should be taken to ensure that the demonstration and instruction at this station match the demonstration and instruction at the other stations. Students should be asked if they have any questions, and answers provided accordingly. Once all questions are answered, the greeter should direct students to the different stations: one to the Field Screening Station, two to the Balance Screening Station, and one to Cone Adaptation Station.

Timeline:
15-20 minutes or as long as necessary to answer questions

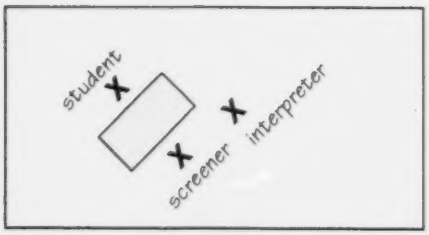
Section III:
The Station Screening

B. Station 1: Cone Adaptation Test

People and Materials Needed:
- Screener (may utilize two screeners if two separate rooms with the same light level are available)
- Interpreters (equal to the number of screeners)

For each screening room:
- Cone Adaptation Test
- Night light
- Table with dark top
- Three chairs
- Two timers or stop watches
- *Cone Adaptation Form* (within student folder)

Room Setup:
Select a screening room with lights that can be turned off but also possessing an additional, dim source of light (e.g., a night light, a light from a door or window, an outside window with blinds closed and possibly covered with sheet or blanket). The dim lighting in the room should imitate dusk or twilight. The source of light must be indirect and not fall upon the screening materials. The student and screener should be seated across from one another at a table. The interpreter should be standing next to the screener, in full view of the student. (See diagram at left.)



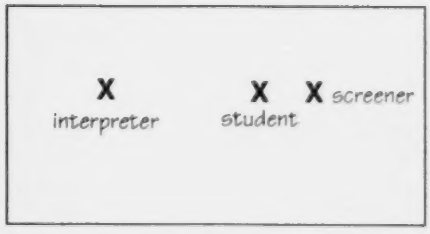
Procedures to be followed by screener:
a) Review procedures on *Cone Adaptation Form* for more detailed instructions.
b) Re-explain the Cone Adaptation Test.
c) Explain that when the light is turned off, the student begins the activity.
d) Conduct the Cone Adaptation Test and document results on the student's Cone Adaptation Form.
e) Reinforce the student for completing the test.
f) Conduct the Cone Adaptation Test and document results again.

g) Reinforce the student for completing the second test. The screener may choose to share the results with the student.
h) Return *Cone Adaptation Form* to the student folder. Give the folder to the student.

Notes:
1. Document on the screening form any irregularities that occur (e.g., squares fall on the floor; the student uses his or her hands instead of both eyes to locate squares).
2. For young children (preschoolers through third grade), explain the activity, give the student an opportunity to practice sorting and stacking the squares, and then conduct the test with the lights off.

Timeline:
Allow 15 minutes per student

C. Station 2: Balance Test
People and Materials Needed:
- Two screeners
- Two interpreters
- *Balance Form* (within student folder)



Room Setup:
Unlike the Cone Adaptation Test, two students can be screened for balance in the same room. Each student should stand facing an interpreter. One screener should stand behind each student. (See diagram at left.)

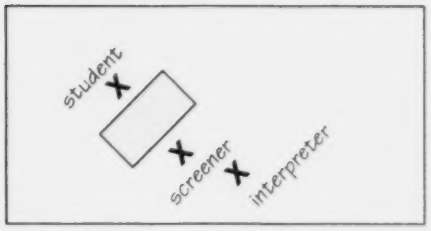
Procedures to be followed by screener:
a) Review procedures on the *Balance Form* for detailed instructions.
b) Re-explain the procedures for the Balance Test.
c) Tell the student to see if he can keep his balance during the first component of the test.
d) Conduct the first component of the Balance Test.
e) Explain that the second component of the test could be more difficult than the first. Ensure the student that you will provide support if he or she loses balance or sways.

Section III:
The Station Screening

f) Conduct the second component of the Balance Test.
g) Reinforce student for completing the test.
h) Document results.
i) Return *Balance Form* to student folder.

Timeline:
Allow 10 minutes per student

D. Station 3: Field Screening
People and Material Needed:
- One screener
- One interpreter
- Three chairs
- *Field Screening Form* (within student folder)



Room Setup:
Both the screener and student should be seated facing each other. There should be approximately one arm's length between them. The interpreter should be standing behind the screener. (See diagram at left.)

Procedures to be followed by screener:
a) Review procedures on the *Visual Field Form* for more detailed instructions.
b) Re-explain the procedures for the Field Screening.
c) Determine which test you will conduct.
d) Explain the activity to the student.
e) Conduct the activity.
f) Reinforce the student for completing the test.
g) Document results.
h) Return the *Field Screening Form* to the student folder.

Note:
If the student has only one eye, make note on the results of the *Field Screening Form.*

Timeline:
Allow 10 minutes to complete the activity.

Section IV:

Evaluating the Results

Once you have completed the paper and station screening, compile data for each student onto an *Usher Screening Data Summary Sheet* (Appendix C). Obtain missing data before proceeding.

Once the form has been completed, there are four possible actions you can take: a) pass the student (i.e., eliminate from further screenings), b) re-check the student within one year, c) refer the student for an ophthalmological evaluation, or d) refer the student for an ERG and visual field testing. The criteria for each of these actions are summarized below.

Pass the student if:

1. Neither biological parent is deaf OR:
   - the family history does not show a history of deafness with a visual impairment,
   - the student began walking at the normal developmental age, OR
   - the student does not show difficulty with balance.
2. The student has been diagnosed with a syndrome other than Usher syndrome or multiple organs were affected at the time of birth that were not the result of trauma, disease, or infection.
3. The student's audiogram does not reflect a corner, a slope, or a slope with a ski bump.
4. The student wears glasses and the family history, student interview, and behavior checklists do not reflect a pattern of visual field loss.
5. The student passed all three station screenings (cone adaptation, balance, and visual field), OR:
   - The student passed the Visual Field Test, passed 75% or better of the Cone Adaptation Test (within a two-minute time period), and passed 75% or better of the Balance Test, OR
   - The student passed the Visual Field Test and Cone Adaptation Test, and 75% of the Balance Test.

If the student meets all of the above criteria, the results indicate a pass. If the family history shows an inconsistent pattern of responses and the behavior checklist indicates possible symptoms of a visual field loss, re-check the student in one year (with both paper and station screenings).

Section IV:

Evaluating the Results

Re-check the student in one year if:

1. The student is under the age of six or had difficulty understanding the testing instructions or demonstration.
2. The family history is unclear as to a past history of hearing loss with a visual impairment, the student did not begin walking at the normal developmental age, or the student has difficulty with balance.
3. The student has multiple organs that were affected after birth due to infection, disease, or trauma.
4. The student's audiogram reflects a corner, a slope, or a slope with a ski bump.
5. The family history, student interview, and behavior checklists are inconsistent with each other in regard to night blindness, a pattern of visual field loss, or other visual losses.
6. The student passed all three station screenings (cone adaptation, balance, and visual field), OR the student passed the Visual Field Test, less than 75% of the Cone Adaptation Test (within a two-minute time period), and less than 75% of the Balance Test.

If the student meets all of the above criteria, the results indicate a re-check. If the family history shows an inconsistent pattern of responses and the behavior checklist indicates possible symptoms of a visual field loss, refer the student for an ERG. (It is better to refer on the side of caution.)

Refer for an ophthalmological evaluation if:

1. The family history does not indicate a history of hearing loss with a visual impairment, the student began walking at the normal developmental age, and the student has difficulty with balance.
2. The student does not have multiple organs affected or the organs were affected at the time of birth.
3. The student's audiogram reflects a corner, a slope, or a slope with a ski bump, OR the audiogram shows a consistent pattern of loss over time.
4. The family history, student interview, and behavior checklists reflect symptoms of acuity loss, blind spots, eye muscle imbalance, glare and contrast sensitivity, or color bindness.
5. The student passed all three station screenings (visual field, cone

Section IV:

Evaluating the Results

adaptation, and balance) OR the student passed the Visual Field Test, more than 75% of the Cone Adaptation Test (within a two-minute time period), and less than 75% of the Balance Test.

If the student meets all of the above criteria, the results of the test indicate a need for an opthalmological evaluation other than an ERG.

Refer for an ERG and extensive visual field evaluation if:

1. The family history indicates both biological parents have Usher syndrome, one parent has Usher syndrome, or a past history of hearing losses with a visual impairment.
2. The student did not begin walking within the normal developmental age and has difficulty with balance.
3. The student does not or does have multiple organs affected or the organs were affected after birth due to trauma, illness, or disease.
4. The student's audiogram reflects a corner, a slope, or a slope with a ski bump.
5. The student behaviors from the family history, student interview, and behavior checklists reflects symptoms of visual field loss and night blindness.
6. The student did not pass the Visual Field Test or was not able to test.
7. The student passed less than 75% of the Cone Adaptation Test (within a two-minute time period).
8. The student passed less than 75% of the Balance Test.

If the student meets all of the above criteria, refer for an ERG and extensive visual field evaluation.

Section V:
References

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Section VI:
Glossary

acuity loss: related to nearsightedness (the ability to see objects close to the body, but not at a distance) or farsightedness (the ability to see objects at a distance, but not close to the body).

ASL: a sign language system that stands for American Sign Language.

assistive listening devices: units that are equipped with a microphone and receiver that helps the individual listen to the speaker while interrupting surrounding conversations and environmental sounds.

audiogram: a chart that indicates what a person does not hear in response to the speed and loudness of sound.

bilateral hearing loss: a hearing loss in both ears.

blind spots: spots that affect the visual field. They may occur as a spot, a series of spots, or in a ring. Blind spots also are referred to as scotomas.

blindness: a person who sees 20/200 with correction and/or who has a 20 degree visual field.

CASE: a sign language system that stands for Conceptually Accurate Signed English.

central acuity loss: a vision loss that occurs in the central part of the retina that reduces the person's central field of vision, acuity, ability to see detail, and color vision.

CHARGE Association: a condition in which the cause is unknown, resulting in colombomas of the eye, heart defects, nasal atresia, delayed growth, genital involvement in males, and ear involvement including deafness.

conductive hearing loss: a loss that occurs in the middle ear usually related to infection, or malfunction of the structures in the middle ear. This loss typically can be corrected with treatment.

cones: the part of the retina that allows the eye to see color.

congenital deafness: a person who is born deaf or hard of hearing.

deafness: not able to hear or detect the production of sounds.

decibels: the measure of intensity or loudness of sound. The notation for decibels is dB.

electroretinogram (ERG): a test conducted by an ophthalmologist that measures the electricity discharged by the nerve impulses in the retina. (For more about ERG, see "What is an Electroretinogram?," page 28.)

geneticist: a doctor who detects genetic conditions and provide genetic counseling.

Goldenhar Syndrome: an inherited condition resulting in irregular facial structure, skin folds located on the lower corner of the eyeball, astigmatism, deafness, and spinal column involvement.

hear of hearing: a person who has a hearing loss and is able to hear or detect some sounds at various decibels and hertz.

hertz: the frequency or rate at which sounds travel. The notation for hertz is hz.

low vision: a person who sees approximately 20/60-20/70 to 20/200 with correction or who has 40 degree visual field. Sometimes referred to as partially sighted.

OD: a notation in a doctor's report that indicates right eye.

ophthalmologist: a medical doctor who assesses and diagnoses visual conditions, and performs medical interventions based on the visual conditions.

OS: a notation in a doctor's report that indicates left eye.

OU: a notation in a doctor's report that indicates both eyes.

Refsum Syndrome: an inherited condition characterized by changes in metabolism resulting in deafness, vision loss, and balance difficulties which are degenerative.

retina: the structure that contains the rods and cones of the eye that allows color vision, detail vision, night vision, and the ability to see darkness affecting the retina. It also affects the blood vessels.

Retinitis Pigmentosa (RP): a progressive, genetic condition that reduces the peripheral field of vision and the ability to see darkness affecting the retina. It also affects the blood vessels. (For more information, see "What is Retinitis Pigmentosa?," page 26.)

rods: the part of the retina that allows the eye to see movement and in the dark.

Rubella: a viral infection that is transmitted to the fetus during pregnancy resulting in cataracts, glaucoma, myopia, hearing loss, heart disease, and mental retardation. The severity of these conditions is dependent upon the time it is acquired.

scotoma: a blind spot or spots in the visual field.

SEE: a sign language system that stands for Signing Exact English.

sensorineural hearing loss: a hearing loss that occurs in the inner ear usually related to malfunction of the structures in the inner ear. The loss typically cannot be corrected with treatment.

unilateral hearing loss: a hearing loss in one ear.

Section VII:
Resources

Written & Visual Resources

Balkwill, F., & Ralph, M. (1995). ***How did I get Usher syndrome?*** London: Harper Collins Publishers.

Information on how Usher syndrome is acquired is presented in a comic book format with an eyeball serving as narrator. The eye takes a young woman through a journey of how she inherited Usher syndrome, and the impact that Usher syndrome has on her activities of daily living.

To obtain a copy:
National Deaf-Blind & Rubella Association
11-13 Clifton Terrace
Finsbury Park, London N4 3SR
Voice: 0171 272 7774; FAX: 0171 272 3862

Davenport, S.L.H. (1996). ***Important topics in deafblind education: Usher syndrome I, II, and III.*** (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University.

This 20-minute videotape presents the characteristics inherent in all three types of Usher syndrome. Typically, students who have Usher syndrome I will experience visual field loss early, are born profoundly deaf, and have poor balance. Students with Usher syndrome II begin losing vision during their late teens, are born with a moderate to profound hearing loss, but do not experience balance problems. Students with Usher syndrome III are very rare. These students have a visual field loss similar to those with Usher syndrome II, are born with normal hearing or have a mild hearing loss that rapidly decreases within a period of 10 years, and may or may not have balance problems. Some preliminary characteristics and suggested modifications to compensate for the vision loss and balance difficulties are described.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Davenport, S.L.H. (1996). ***Important topics in deaf-blind education: Patterns of inheritance.*** (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University.

This 12-minute videotape presents the basics of genetics and traits inherited by males and females. Recessive, dominant, and X-linked characteristics are passed through generations. These traits have significant impact on the physical body of individuals in various forms. Several conditions are presented and discussed, such as Tay Sachs, Stickler syndrome, Treacher Collins syndrome, PKU, and Usher. Many of these conditions affect both vision and hearing sensory systems.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Davenport, S.L.H. (1997). ***Important topics in deafblind education: Retinitis Pigmentosa in Usher syndrome.*** (videotape). Terre Haute: Blumberg Center for Inter-disciplinary Studies in Education, Indiana State University.

This 20-minute videotape presents the impact Retinitis Pigmentosa has on vision. A description of the progressive nature of the syndrome is presented. Actual pictures of a retina affected by retinitis pigmentosa are displayed. Additionally, behaviors that may indicate early signs of retinitis pigmentosa are identified including night blindness and tunnel vision.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Davenport, S.L.H. (1996). ***Important topics in deafblind education: Balance.*** (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University.

This 20-minute videotape illustrates anatomical functions that control balance including vision, muscle structure, hea-ing, the cerebellum, and joints. It takes the viewer through a journey of child motor development. This video also includes clips of children with various stages of CHARGE Association showing their chronological ages and their histories of motor and balance development.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Davenport, S.L.H. (1996). ***Important topics in deafblind education: Genetics and causes of birth defects*** (videotape). Terre Haute: Blumberg Center for Interdisciplinary Studies in Education, Indiana State University.

This 16-minute videotape presents conditions that can impact the genetic system during embryonic development including multifactorial elements, infections, narcotics and alcohol, diet, chromosomal, and the environment. Descriptions of steps a geneticist conducts to determine the location and type of syndrome are presented. Some infections related to deafblindness are described, such as Toxoplasmosis, Rubella, Cytomeglovirus, Herpes, and Syphilis (TORCH).

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Edwards, L., Goehl, K.S., Gordon, L., & Houghton, J. (1992). ***Profiles: Individuals with deaf-blindness.*** Terre Haute: Indiana Deaf-Blind Services Project, Indiana State University.

This manual provides descriptions of common syndromes related to deaf-blindness. Each syndrome listed contains information on incidence, impact of the vision and hearing loss, physical descriptions, and personal testimonies of individuals who have the condition. This is a very powerful and informative manual that brings in the human component as well as the medical one.

To obtain a copy:
Karen Goehl
Blumberg Center for Interdisciplinary Studies in Education
SOE 502
Indiana State University
Terre Haute, IN 47809
Voice: (812) 237-2830

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Section VII:
Resources

Enos, J. & Jordan, B. (1996) ***A guide for students who are deaf-blind considering college.*** Sands Point, NY: Helen Keller National Center-Technical Assistance Center.

This guide was developed for students who are pursuing education beyond high school and for people assisting these students achieve their goals. The document helps students assess their own learning styles so they can better advocate for their needs. In addition, it helps them assess the strengths and weaknesses of potential post-secondary settings.

To obtain a copy:
Kathy Michaels
National Technical Assistance Consortium
111 Middle Neck Road
Sands Point, NY 11050
Voice: (516) 944-8900 x307
TTY: (516) 883-9059; FAX: 516-883-9060

Gallaudet College. (1978). ***A screening program for Usher's syndrome.*** Washington, DC: Author.

This manual presents a vision screening model for Usher syndrome that educationally assesses the person's visual field only. Other data gathering tools (e.g., family history, behavioral characteristics, and student interviews) are listed as necessary to complete screening activities. Although most screening models consist of additional types of evaluations related to sight, many of the suggestions and ideas in this manual are pertinent to screening sites.

To obtain a copy:
Public Service Programs, Deaf-Blind Program
Gallaudet University
800 Florida Ave. NE
Washington, DC 20002
Voice: (202) 651-5051; TTY: (202) 651-5052
FAX: 202-651-5054

Hennessey, J.C. (1991). ***The inheritance of RP and allied retinal degenerative diseases.*** Baltimore: National Retinitis Pigmentosa Foundation, Inc.

This booklet presents the genetic pattern of retinitis pigmentosa and the patterns of inheritance. Autosomal dominant, autosomal recessive, and sex-linked transmissions are described. Retinitis Pigmentosa can occur in any one of the above listed patterns. A glossary of terms makes this document easy to read and understand.

To obtain a copy:
National Retinitis Pigmentosa Found., Inc.
d/b/a RP Foundation Fighting Blindness
1201 Mt. Royal Avenue, 4th Floor
Baltimore, Maryland 21217
Toll free: 1-800-683-5555
Voice: (410) 225-9400; TTY: (410) 225-9409
FAX: 410-225-3936

Indiana Deaf-Blind Services Project. (1997, draft). ***What is Usher syndrome: How to recognize the combination of hearing loss and retinitis pigmentosa.*** Terre Haute: Blumberg Center for Interdisciplinary Studies, Indiana State University.

This manual describes conditions related to Usher syndrome using a question and answer format. It provides information related to behaviors often related to individuals with Usher syndrome as vision loss progresses. It discusses the importance of early detection, with descriptions of screening tests and ophthalmologic testing. Resources for learning more about Usher syndrome are located at the end of the manual.

To obtain a copy:
Karen Goehl
Blumberg Center for Interdisciplinary Studies in Education
SOE 502
Indiana State University
Terre Haute, IN 47809
Voice: (812) 237-2830

Kansas Usher Screening Committee. (1996). ***A collection of resources on Usher syndrome.*** Topeka: Author.

This packet contains information on Usher syndrome from various state and national organizations. Additonally, there are two resource lists of specialists in the greater Wichita and Kansas City areas focusing on ophthalmological evaluations, genetic counseling, support services, and social-emotional counseling.

To obtain a copy:
Kansas Dual Sensory Impairments Project
Kansas State Department of Education
120 SE 10th Ave.
Topeka, Kansas 66612
(785) 296-0917 (V/TTY)

Helen Keller National Center Regional
4330 Shawnee Mission Parkway, Ste. 108
Shawnee Mission, Kansas 66205
(913) 677-4562 (V/TTY)

Kansas State School for the Deaf
Department of Nursing
450 East Park
Olathe, Kansas 66061
(913) 791-0573

Kansas State School for the Blind
Outreach Department
1100 State Ave.
Kansas City, Kansas 66102
(913) 281-3308 (Voice)

Section VII:
Resources

McGann, R. (1995). ***Tactile interpreters for students who are deaf and blind.*** (videotape). Gibsonia, PA: Distance Learning Center.

This two-hour videotape presents various strategies to use when communicating with someone who is deaf-blind including those individuals with Usher syndrome. The presenter demonstrates how to approach a person who is deaf-blind, assist a person who is deaf-blind, and end a conversation with a person who is deaf-blind. Tactile sign language is introduced, including components of this communication system. This video is accompanied by a booklet that highlights important aspects presented in the tape.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Miner, I., &, Cioffi, J. (1997). ***Usher syndrome in the school setting.*** Sands Point, NY: Helen Keller National Center-Technical Assistance Center.

This resource presents issues around the identification, intervention, and supports needed for students with Usher syndrome. The authors stress that early intervention and contact with adults who have Usher syndrome are necessary if students are to feel good about themselves and have a sense of their futures. Strategies that assist in developing good self-esteem and essential life skills consist of participating in problem solving, sound decision-making, and introducing role models. Introduction to these strategies should occur early in the student's education as they can enhance successful life experiences as the disability progresses. This manual lists next steps, national and state resources, information and support regarding the education and rehabilitation of students with Usher syndrome.

To obtain a copy:
Kathy Michaels
National Technical Assistance Consortium
Helen Keller National Center
111 Middle Neck Road
Sands Point, New York 11050
Voice: (516) 944-8900 x 307
TTY: (516) 883-9059; FAX: 516-883-9060

Sims, B., Raistrick, K., McGann, R., Schmidlin, R., Iurlano, A., & Baumgarner, J. (1996, March). ***When hearing loss and Retinitis Pigmentosa happen together: Meeting educational needs.*** (videotape). PennTech Teleconference Presentation, Gibsonia, PA: Educational Service Center.

This three-hour video describes the symptoms for each type of Usher syndrome. Individuals with Usher syndrome talk about their experiences growing up with the condition. Accommodations and modifications needed when there is a decrease in the visual field are presented. National resources are described. A paper highlighting the issues discussed in the videotape accompanies the tape.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Rothstein, R. (1998). ***The light at the end of the tunnel: Practical strategies for living with Usher syndrome.*** Sands Point, NY: Helen Keller National Center.

This book is written for individuals with Usher syndrome, but will also be helpful for relatives, friends, teachers, counselors, employers, co-workers, and service providers. The first section of the text explains Usher syndrome and its effect on individuals, including vision, hearing, balance, and genetics. The rest of the text looks at how to cope with Usher, including psycho-social and practical methodologies. Much of the information has been gathered from people who have Usher syndrome. The publication also has a resource list of agencies, organizations, and information sources.

To obtain a copy:
Barbara Hausman
Helen Keller National Center
111 Middle Neck Road
Sands Point, NY 11050
Voice: (516) 944-8900; TTY: (516) 944-8637
FAX: 516-944-7302

Smith, T. (1992). ***Deaf-blind communication and community: Overview and introduction.*** (videotape). Burtonsville, MD: Sign Media Inc.

This one-hour, open-captioned videotape discusses a number of topics relevant to communication for individuals who are deaf-blind with an emphasis in communication with individuals who have Usher syndrome. It contains clips of various individuals participating, traveling, and communicating within their community environments.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

Smith, T. (1992). ***Deaf-blind communication and community getting involved: A conversation.*** (videotape). Burtonsville, MD: Sign Media Inc.

This 90-minute videotape presents live conversations with individuals who have Usher syndrome. They discuss how they learned they had Usher syndrome, changes they had to make in their lives once their vision deteriorated, and career opportunities. These individuals discuss the necessary supports and supplementary aids that assist individuals with Usher syndrome. Interpreters also share some of their experiences in working with individuals who have Usher syndrome.

To borrow a copy:
Kansas Deaf-Blind Loan Library,
KIRC, Kansas City, Kansas
1-800-572-5463, ext. 5

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Section VII:
Resources

National Resource Agencies

American Association of the Deaf-Blind (AADB)
814 Thayer Avenue, Suite 302
Silver Springs, Maryland 20910-4500
TTY: (301) 588-6545; FAX: 301-588-8705
E-mail: aadb@erols.com

Boys Town National Research Hospital
Genetics Department
555 N. 30th Street
Omaha, Nebraska 68131
Voice/TTY: (800) 835-1468
FAX: 402-498-6331

Dr. Sandra Davenport
Sensory Genetics/Neuro-development
5801 Southwood Drive
Bloomington, Minnesota 55437-1739
Voice/TTY: (612) 831-5522

Helen Keller National Center (HKNC)
111 Middle Neck Road
Sands Point, New York 11050
Voice: (516) 944-8900
TTY: (516) 944-8637; FAX: 516-944-7302
Webpage: http://www.helenkeller.org

National Technical Assistance Consortium Headquarters (NTAC)
Teaching Research Division
Western Oregon University
345 N. Monmouth Avenue
Monmouth, Oregon 97361
Voice: (503) 838-8807
TTY: (503) 838-9623; FAX: 503-838-8150
Webpage: http://www.tr.wou.edu

DB-Link
Teaching Research Division
Western Oregon University
345 N. Monmouth Avenue
Monmouth, Oregon 97361
Voice: 1-800-438-9376
TTY: 1-800-854-7013
Webpage: http://www.tr.wou.edu/dblink

Retinitis Pigmentosa Foundation
1401 Mount Royal Avenue, 4th Floor
Baltimore, Maryland 21217-4245
Voice: (410) 225-9400; FAX: 410-225-3936

National Information for the Center on Deafness (NICD)
Gallaudet University
800 Florida Avenue NE
Washington, District of Columbia 20002
Voice: (202) 651-5051
TTY: (202) 651-5052; FAX: 202-651-5054

Usher Around the World
c/o Vision Screening Project
5801 Southwood Drive
Bloomington, Minnesota 55437
Voice/TTY: (612) 831-5522
FAX: 612-831-0381

Fighting Blindness News
The Foundation for Fighting Blindness
National Retinitis Pigmentosa Foundation, Inc.
Executive Plaza 1, Suite 800
11350 McCormick Road
Hunt Valley, Maryland 21031-1014
Voice: (800) 638-5555
Webpage: http://www.blindness.org

Hereditary Deafness Newsletter of America
The National Research Register for Hereditary Hearing Loss
Boys Town National Research Hospital
555 N. 30th Street
Omaha, Nebraska 68131
Voice/TTY: (402) 498-6631
Voice: (402) 498-6739

State & Regional Resources

Helen Keller National Center, Great Plains Regional Office
4330 Shawnee Mission Parkway, Ste. 108
Shawnee Mission, Kansas 66205
Voice/TTY: (913) 677-4562
FAX: 913-677-1544
E-mail: HKNC7BJ@sprintmail.com

National Technical Assistance Consortium (NTAC) Regional Office
4330 Shawnee Mission Parkway, Ste. 108
Shawnee Mission, Kansas 66205
Voice/TTY: (913) 677-4562
FAX: 913-677-1544
E-mail: 76434.2431@compuserve.com

National Family Association for Deaf-Blind
Great Plains Region VII
630 W. 68th Terrace
Kansas City, Missouri 64113
Voice: (816) 333-8459; FAX: 816-333-3369
E-mail: Kurt.Kavanaugh@MCIONE.com

Kansas Dual Sensory Impairments Project
Kansas State Department of Education
120 SE 10th Avenue
Topeka, Kansas 66612
Voice/TTY: (785) 296-0917
FAX: 785-296-1413
E-mail: jhoughton@ksbe.state.ks.us

Marta Vinton
Usher Support Group
8331 W. 108th St., Apt. H
Overland Park, Kansas 66212
TTY: (913) 338-5288; FAX: 913-6631276

David Bennett, Affiliate
Helen Keller National Center
Envision Inc.
801 E. Lincoln
Wichita, Kansas 67211

White Canes & More
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, Kansas 67214
Voice: (316) 681-0870
FAX: 316-682-4747

Section VII:
Resources

Counseling

Cindy Winsky, State Coordinator for the Deaf
David Tout, Case Manager & Therapist
Kelly Migues, Case Manager & Therapist
Johnson County Mental Health Center
1125 W. Spruce
Olathe, Kansas 66061
Voice/TTY: (913) 782-2100

Ron Lybarger, Therapist
6142 Kenwood Avenue
Kansas City, Missouri 64110-3348
Voice/TTY: (816) 822-3679

Jan Rodgers, Family Therapist
Family Institute of Kansas City, Inc.
9250 Ward Parkway, Suite 105
Kansas City, Missouri 64114
Voice: (816) 822-1717 x2

Tag Goodspeed, Therapist
High Plains Mental Health Center
208 E. 7th St.
Hays, Kansas 67601
Voice: (785) 628-2871

John Kingsley, Therapist
ComCare of Sedgwick County
1919 N. Amidon St., Suite 130
Wichita, Kansas 67203
Voice: (316) 832-0318
TTY: (316) 832-1813

Ophthalmological Evaluations

Dr. Trudi Grin, Ophthalmologist
10550 Quivira Road
Overland Park, Kansas 66215
Voice: (913) 888-1888

Dr. Gerhard Cibis, Pediatric Ophthalmologist
4620 JC Nichols Parkway
Kansas City, Missouri 64112
Voice: (816) 444-2943

University of Kansas Medical Center
Department of Ophthalmology
3901 Rainbow Boulevard
Kansas City, Kansas 66160
Voice: (913) 588-6600

Children's Mercy Hospital
Department of Opthalmology
2401 Gillham Road
Kansas City, Missouri 64108
Voice: (816) 234-3000

Geri McFadden, Coordinator
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, Kansas 67214
Voice: (316) 682-4646
FAX: 316-682-4747

Dr. Linda Buettner
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, Kansas 67214
Voice: (316) 682-4646
FAX: 316-682-4747

Dr. Leslie Nesmith, Opthalmologist
530 N. Lorraine
Wichita, Kansas 67214
Voice: (316) 683-5611
FAX: 316-683-0294

Dr. Michael Varenhorst, Opthalmologist, Retina Specialist
530 N. Lorraine
Wichita, Kansas 67214
Voice: (316) 682-4646
FAX: 316-682-4747

Dr. David Waldie, OD,
Director of Optometry
Vision Rehabilitation Center
530 N. Lorraine, Suite 100
Wichita, Kansas 67214
Voice: (316) 682-4646
FAX: 316-682-4747

Dr. Paul Wieshaar
Opthalmologist, Retina Specialist
530 N. Lorraine, Suite 100
Wichita, Kansas 67214
Voice: (316) 682-4646
FAX: 316-682-4747

Genetic Counseling

Dr. David Harris, Geneticist
Children's Mercy Hospital
2401 Gillham Road
Kansas City, Missouri 64108
Voice: (816) 234-3290

Dr. Sechin Cho, Geneticist
Wesley Medical Center
550 N. Hillside
Wichita, Kansas 67214
Voice: (316) 688-2080

Equipment Resources

Cone Adaptation Test
Precision Vision
745 N. Howard
Villa Park, IL 60181
Voice: (630) 833-1454
FAX: 630-833-1520

What's Tunnel Vision? (simulators)
RP Research Foundation
366 Adelaide Street West, Suite 704
Toronto, Ontario, Canada M5V 1R9
Voice: (416) 598-4951

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Appendix A:

What Is Retinitis Pigmentosa?

Retinitis Pigmentosa (RP) is an inherited condition. It affects the sensory cells (rods and cones) and the blood vessels of the retina. It may cause pigmentary clumping and a loss of electrical responses in the area of the eye. Usually the peripheral retina is affected first. This area of the retina contains the largest majority of rods (150 million) which allows a person to see in dim light, have night vision, and detect outer movement. Once the rods are affected, a person may have spotty vision (scotomas), a ring of missing information (ring scotoma), or tunnel vision (outer, lower, and upper fields are decreased).

The cones are affected next. The majority of cones are located in the macular (central) part of the retina. The fovea, a spot of the retina that contains the greatest number of cones (7 million), does not contain rods. The cones allow a person to see color, fine detail, and printed words on a page. By the time the cones are affected, there are jagged spots, the pigment separates forming clumps of pigment, bone spicules, dark spots that can be seen in the inner structures of the eye, and the optic disk is pale, yellow, and waxy. The macula may become swollen and cone cells in the fovea may die. A person may have difficulty viewing objects that are colored, small print, objects presented in the central part of the visual field, or fine detail.

Another condition that may be present which will affect central vision (Usher Type I) are cataracts. These may develop in the lens of the eye and may be present by the time a person reaches 20-40 years of age. Once cataracts have developed, a person may have difficult seeing bright light, fine detail, and printed words. Everything may appear cloudy.

Appendix A:
Characteristics of Usher Syndrome and Sensorineural Hearing Loss

Different Types of Sensorineural Hearing Loss

TYPE	DECIBLE RANGE	WHAT CAN A PERSON HEAR?
Normal	0-15 dB	Everything: all environmental sounds and speech sounds
Mild to moderate	15-30 dB	Watch ticking, whispers, & waterfall; sounds of f, th, s, z
Moderate	30-50 dB	Normal conversational speech but not the full spectrum of sounds; j, m, d, b, n, ng, e, i, a, o, r, p, h, g, ch, sh, k
Moderate to severe	50-70 dB	Air conditioner; loud voices; difficulty in group situations
Severe	70-90 dB	Piano, food blender, loud vowel sounds, lawn mower, telephone, dog barking
Profound	90-110 dB	Chain saw, semi truck, helicopter, train, airplane, very loud horn

Characteristics Associated with the Three Types of Usher Syndrome

	TYPE I	TYPE II	TYPE III
HEARING LOSS	Congenital severe to profound bilateral sensorineural hearing loss; corner audiogram	Congenital moderate to profound bilateral sensorineural hearing loss; sloping audiogram	Normal hearing to moderate bilateral sensorineural hearing loss at birth; rapid progression of hearing loss for 5-10 years; sloping audiogram with a ski bump
VISION LOSS	Early onset of blind spots; early onset of night blindness; peripheral vision loss; usually results in total blindness during adult years.	Blind spots or ring scotoma by teenage years; peripheral vision loss; usually does not progress to total vision loss	Blind spots or ring scotoma by teenage years; peripheral vision loss; progression varies but usually legally blind by young adult years
OTHER VISUALLY-RELATED CONDITIONS	Early development of cataracts	Varied central acuity loss	Progression varies but nighttime vision loss precedes daytime vision loss
BALANCE	Balance problems; walks at 18-24 months; does not get dizzy	Balance normal	May have some balance problems
GENETICS	1A long arm of 14; 1B long arm of 14 (most common); short arm of 11 (Acadian descent)	2A long arm; 2B long unknown	3A long arm of 3 (Finnish descent)

Appendix A:

What Is an Electroretinogram (ERG) Evaluation?

An ERG is an evaluation that measures the electricity discharged by nerve impulses in the retina. This test is conducted by a board certified ophthalmologist. It can predict RP in children beginning at six to seven years old and for some children it can be predicted at younger ages (reportedly as young as three years old).

This test is conducted using two methods: the skin electrode method and the lens method. When using the skin electrode method, a person has electrodes attached to the skin in certain locations around the eye. These electrodes are connected to a computer which will record responses of the retina. The person places his or her head in a globe called a Gonzfield Globe. Flashes of colored lights are presented in various locations in the globe for a brief amount of time.

When using the lens method, a person wears contact lenses with electrodes attached. These electrodes are connected to a computer. The person is presented with flashes of colored light in the globe, like the one used in the skin electrode method. As in the skin electrode method, the responses made by the retina are recorded. The doctor will interpret the responses that indicate onset of RP.

Appendix A:

Usher Screening Inservice Agenda

Usher Screening Inservice Agenda
(names of presenters/coordinator of agenda)
(date)
(location—at prospective screening site)

15 min	Welcome & Introduction of Screening Team Members
30 min	What Is Usher Syndrome?
10 min	What Are We Doing in Kansas?

Screening for Usher Syndrome

20 min	Run Through a Typical Screening Day
10 min	Role Assignments
	Match Screeners to Stations
	How to Work with an Interpreter
20 min	Determine Station Locations
25 min	Role-Play Station
25 min	Cone Adaptation Station
25 min	Balance Station
25 min	Field Screening Station
5 min	Screening Expectations
5 min	Wrap-Up

Appendix A:

How to Explain Usher Syndrome to Kids

Step 1: Briefly describe what happens with Usher syndrome

"Usher syndrome is when the part of your eye called the retina gets sick. When it gets sick, you cannot see things as well as when you were younger. Your sight may get even worse as you get older.

"If you think your eyes are not seeing well, you need to tell your parents, your teacher, or an adult friend. While you are in school, you may be asked to do some activities with a teacher or nurse that will test how well you see. This is called an Usher Screening Test. There are three parts to this test."

Step 2: Explain the Cone Adaptation Test

"The first is the Cone Adaptation Test. In the Cone Adaptation Test, you will have about 15 squares. Some are white, some are blue, and some are red. An adult will ask you to put all the red squares in one pile, all the white squares in another pile, and all the blue squares in a third pile. You will do this two times. There will be very little light in the room.

"When the lights are off, the adult can learn how your eyes use a little light to help you see. Some students have problems when there is only a little bit of light in the room. If you think you have trouble seeing with just a little light, do not worry. That does not mean you have Usher syndrome."

Step 3: Explain the Balance Test

"The second test is the Balance Test. This test is done with the lights ON in the room. Sometimes your eyes will be open; sometimes they will be closed.

"First, you will stand with ONE FOOT IN FRONT OF THE OTHER. Your hands will touch your legs and your eyes will be open. You will lift your arms up to the side like "you wanted to fly." The adult will stand behind you and lightly touch the sides of your body (trunk) to see if you can remain standing. Then, you will do the same thing with your eyes closed.

"Next you will stand WITH YOUR FEET TOGETHER. Your hands

Appendix A:

How to Explain Usher Syndrome to Kids

will touch your legs and your eyes will be open. You will lift your arms up to the side like "you wanted to fly." The adult will stand behind you and lightly touch the sides of your body to see if you can remain standing. Then, you will do the same thing with your eyes closed. That's it!"

Step 4: Explain the Field Test

"During the Field Test, you will sit directly across from an adult. You will be asked to sit straight and to keep your eyes on the adult's nose at all times. The adult's hands will be at the side of his or her body. When you see the adult's fingers wiggle, raise your hand. You will do this two or three times, then you'll be finished! The important thing is to keep your eyes on the adult's nose. Don't worry if you might have difficulty seeing the wiggling fingers. This does not mean you have Usher Syndrome.

"Remember, it is important to ALWAYS ASK QUESTIONS. If you do not understand what to do, are scared about the test, or worried about how you did, tell the adult. Someone will be glad to help you."

Appendix B:

A Case Study/Usher Syndrome I

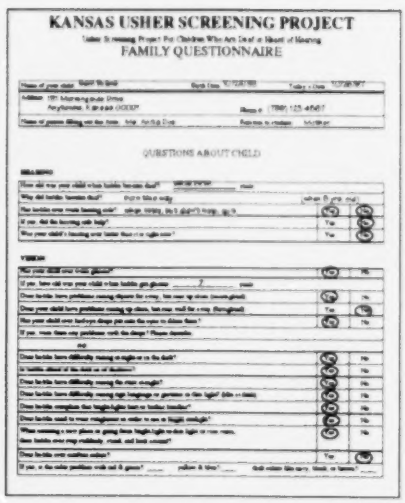
Background: Sam Evans

Sam Evans is 14 years old. He was born with a profound bilateral hearing loss. Sam uses ASL and print for receptive and expressive communication. He possibly has some difficulties with vision, as noted by his teachers and parents. Sam was referred to his school nurse for educational Usher Screening.

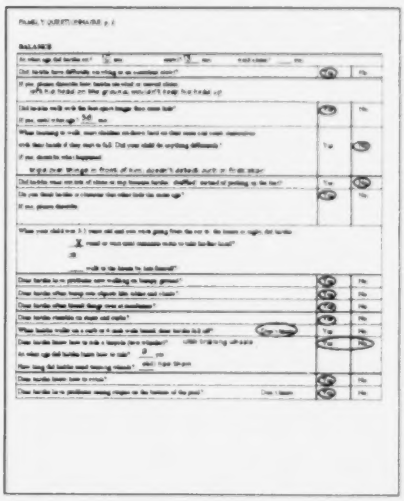
In preparation for the station screening, the nurse sent the family history information to Sam's parents, behavior checklists to all of Sam's teachers, obtained his audiogram from the audiologist, and had Sam's homeroom teacher conduct a student interview with Sam. The results are as follows:

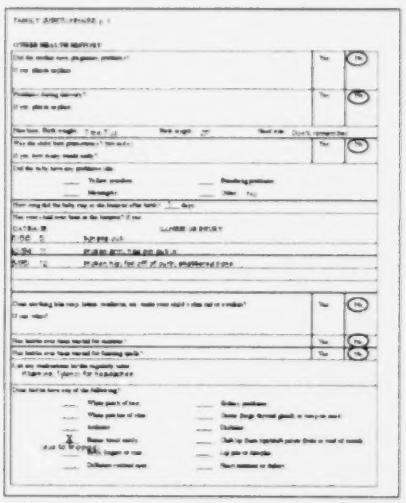
Paper Screening: Family History

Sam's mother completed the form. She indicated that Sam was deaf at birth. He tried hearing aids when he was five, but they were discontinued when they did not appear to help him. She indicated that he had worn glasses since the age of seven. Additionally, she marked several behaviors that indicated Sam was having difficulty seeing at night (e.g., difficulty seeing at night, afraid of shadows, and difficulty seeing signs or gestures in dim light) and difficulty with light adjustment (e.g., wears sunglasses and stops when exiting a darkened room to the outdoors).


KANSAS USHER SCREENING PROJECT

FAMILY QUESTIONNAIRE





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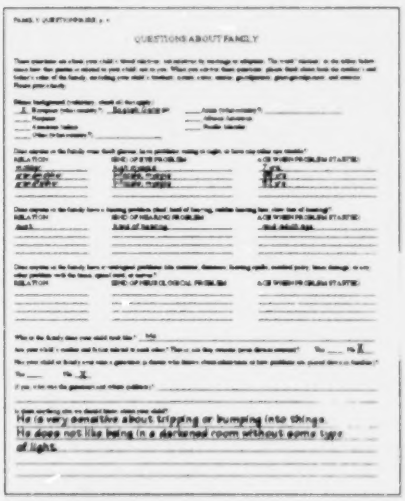
Appendix B:

A Case Study/Usher Syndrome I

She further reported that Sam:

- did not walk until he was 20 months old,
- kept his head on the ground when he crawled,
- trips over things in front of him,
- doesn't see the curb or the first stair of a stairway,
- holds onto someone's hand when the lights are dim or it is dark,
- had difficulty riding a bike,
- was hospitalized for a tonsilectomy, a broken arm, and a broken hip,
- takes vitamins and Tylenol (as needed) for headaches,
- is very sensitive about tipping or bumping into things, and
- does not like to be in a darkened room without the lights on.



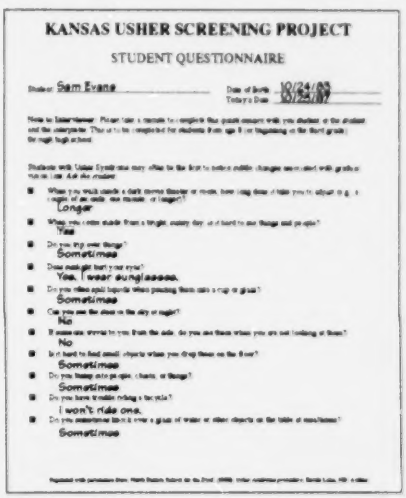
QUESTIONS ABOUT FAMILY

He is very sensitive about tripping or bumping into things.
He does not like being in a darkened room without some type of light.

Although Sam's mother reported no problems during pregnancy, she did indicate a family history of myopia, bifocals, and an aunt who was diagnosed as hard of hearing during "middle age." Neither Sam nor his family members have seen a geneticist.

Paper Screening: Student Interview

Sam reported that his eyes take longer than a minute to adjust to a darkened room. He stated that he sometimes trips over things, spills liquids when pouring from a container into a cup, and has difficulty finding small objects dropped on the floor. He indicated that sunlight bothers his eyes. He wears sunglasses when he is outdoors.



KANSAS USHER SCREENING PROJECT

STUDENT QUESTIONNAIRE

Student: Sam Evans — Date of Birth: 10/24/83 — Today's Date: 05/25/97

- When you walk outside a dark movie theater or room, how long does it take you to adjust (e.g., a couple of minutes, one minute, or longer)? Longer
- When you come inside from a bright, sunny day, is it hard to see things and people? Yes
- Do you trip over things? Sometimes
- Does sunlight hurt your eyes? Yes, I wear sunglasses.
- Do you miss spilling liquids when pouring them into a cup or glass? Sometimes
- Can you see the stars in the sky at night? No
- If someone stands to you from the side, do you see them when you are not looking at them? No
- Is it hard to find small objects when you drop them on the floor? Sometimes
- Do you bump into people, chairs, or things? Sometimes
- Do you have trouble riding a bicycle? I won't ride one.
- Do you sometimes bump a cup of water or other objects on the table or countertop? Sometimes

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Appendix B:

A Case Study/Usher Syndrome I

Paper Screening: Behavior Checklist

Both Sam's mother and his social science teacher completed behavior checklists. There was a consistent pattern in night blindness, visual fields, balance, and other areas. There were inconsistencies reported for glare, contrast, and acuity.

BEHAVIOR QUESTIONNAIRE

Name of child: Sam Evans

Birth date: 10/24/83 Current date: 10/25/97

Completed by: Mr Speaker (social science teacher)

Night blindness

X Can't see when coming in from bright sunlight

X trips over things when light changes or light is dim

X stays near light in a darkened room or at night

____ points out or light falls on the face of a speaker

____ may express a desire to enter a room before it is darkened (e.g. movie theater)

____ avoids conversations in darkened area

X may appear to stagger or lose balance after an exercising car has passed at night

X has problems reading under some lights or in dimly lit areas

Visual Field

X stumbles on stairs and curbs

X bumps into people, tables, and chairs

X may bump or tip over objects placed to the side of most areas

X startles easily

____ seems to hold objects in unusual positions when looking at them

X turns head while reading across a page

X uses fingers to mark place while reading

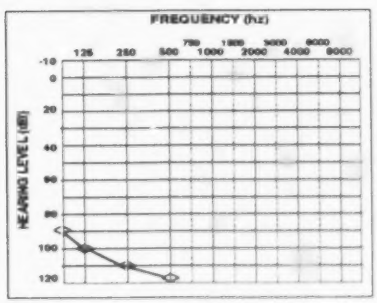
X can't find small objects that have been dropped

X likes to glance at another person's hand waving from the side

X or uses or may edge to one side when in a large group

X frequently trips or falls in unfamiliar or unfamiliar surroundings

Paper Screening: Audiogram



Sam's audiogram revealed a corner audiogram consistent with audiograms that would indicate Usher syndrome I. He has a 90 dB loss that drops off dramatically in the lower frequencies.

Based on the patterns of visual behaviors, his audiogram, and family history, it was determined that Sam needed to receive station screening.

Cone Adaptation Station Screening

Sam completed both trials of the Cone Adaptation Test within 30-45 seconds. He received 100% correct for grouping the white squares, and less than 75% for grouping the red and blue squares.

KANSAS USHER SCREENING PROJECT

Student's Name: Sam Evans Age: 14

Date: 10/25/97

CONE ADAPTATION

Procedure

Results

Trial #1		Trial #2	
2:00	Time started	2:00	Time started
1:30	Timed stopped	1:15	Timed stopped
30 min	Total time	45 min	Total time
4/4 2/4 2/4	Number correct	4/4 1/4 1/4	Number correct
(white) (blue) (red)		(white) (blue) (red)	

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Appendix B:

A Case Study/Usher Syndrome I

Balance Station Screening

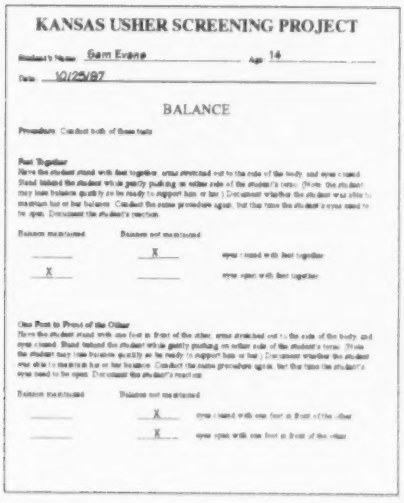
Sam failed 75% of the Balance Test. He only passed the section with his eyes open and both feet together.

Visual Field Station Screening

Sam's visual fields were tested using both eyes. Both horizontal and vertical fields indicated a possible loss in the periphery.

Analysis of station screening:

Since Sam did not pass any of the station screening, his paper screening indicated visual field losses, balance difficulties, night blindness, and his family history indicated the presence of Usher syndrome, the school nurse referred Sam for an ERG and extensive ophthalmological evaluation with a physician with experience in visual field losses related to syndromes. The test was paid for by Sam's parents' insurance. Sam's mother currently is waiting for the test results.



KANSAS USHER SCREENING PROJECT

Student's Name Sam Evans Age 14

Date 10/25/97

BALANCE

Procedure: Conduct both of these tests

Feet Together

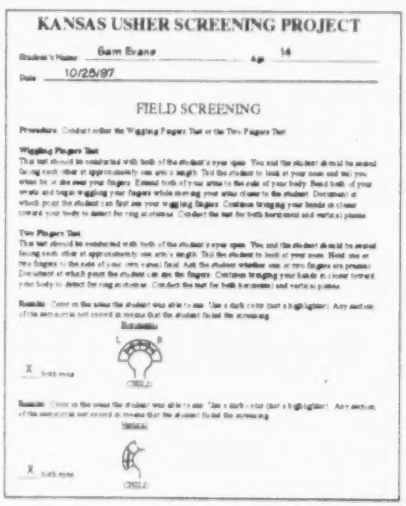
Have the student stand with feet together, arms stretched out to the side of the body and eyes closed. Stand behind the student while gently pushing on either side of the student's torso. (Note: the student may lose balance quickly so be ready to support him or her.) Document whether the student was able to maintain his or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained	Balance not maintained	
_____	X	eyes closed with feet together
X	_____	eyes open with feet together

One Foot in Front of the Other

Have the student stand with one foot in front of the other, arms stretched out to the side of the body and eyes closed. Stand behind the student while gently pushing on either side of the student's torso. (Note: the student may lose balance quickly so be ready to support him or her.) Document whether the student was able to maintain his or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained	Balance not maintained	
_____	X	eyes closed with one foot in front of the other
_____	X	eyes open with one foot in front of the other



KANSAS USHER SCREENING PROJECT

Student's Name Sam Evans Age 14

Date 10/25/97

FIELD SCREENING

Procedure: Conduct either the Wiggling Fingers Test or the Two Fingers Test

Wiggling Fingers Test

This test should be conducted with both of the student's eyes open. You and the student should be seated facing each other at approximately one arm's length. Tell the student to look at your nose and tell you when he or she sees your fingers. Extend both of your arms to the side of your body. Bend both of your wrists and begin wiggling your fingers while moving your arms closer to the student. Document at which point the student can first see your wiggling fingers. Continue bringing your hands in closer toward your body to detect for ring at whatever. Conduct the test for both horizontal and vertical planes.

Two Fingers Test

This test should be conducted with both of the student's eyes open. You and the student should be seated facing each other at approximately one arm's length. Tell the student to look at your nose. Hold one or two fingers to the side of your own visual field. Ask the student whether one or two fingers are present. Document at which point the student can see the fingers. Continue bringing your hands in closer toward your body to detect for ring at whatever. Conduct the test for both horizontal and vertical planes.

Results: Enter in the areas the student was able to see (use a dark color (not a highlighter). Any section of the semicircle not covered in means that the student failed the screening.

Right eyeball

L R

X both eyes

FIELD

Results: Enter in the areas the student was able to see (use a dark color (not a highlighter). Any section of the semicircle not covered in means that the student failed the screening.

Left eyeball

X both eyes

FIELD

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Appendix C:
Blank Forms

Usher Screening Forms (Summary Sheet)

Family Questionnaire

Student Questionnaire

Behavior Questionnaire

Cone Adaptation Test

Field Screening Test

Balance Test

Usher Screening Data Summary

Note: the user of this manual is granted permission to copy these forms.

KANSAS USHER SCREENING PROJECT

Student's Name: _____ **Birth Date:** _____

USHER SCREENING FORMS

Dates

1. Family Questionnaire
   - completed _____
   - audiogram attached (if applicable) _____
2. Student Questionnaire _____
3. Behavior Observations
   - teacher _____
   - parent _____
   - dorm teacher _____
4. Cone Adaptation _____
5. Field Screening _____
6. Balance Screening _____
7. Additional Comments _____

KANSAS USHER SCREENING PROJECT

Usher Screening Project For Children Who Are Deaf or Heard of Hearing

FAMILY QUESTIONNAIRE

Name of your child	Birth Date	Today's Date
Address		
	Phone #	
Name of person filling out this form:	Relation to student:	

QUESTIONS ABOUT CHILD

HEARING

How old was your child when he/she became deaf? _____ years		
Why did he/she become deaf?		
Has he/she ever worn hearing aids?	Yes	No
If yes, did the hearing aids help?	Yes	No
Was your child's hearing ever better than it is right now?	Yes	No

VISION

Has your child ever worn glasses?	Yes	No
If yes, how old was your child when he/she got glasses? _____ years		
Does he/she have problems seeing objects far away, but sees up close (nearsighted)?	Yes	No
Does your child have problems seeing up close, but sees well far away (farsighted)?	Yes	No
Has your child ever had eye drops put into the eyes to dilate them?	Yes	No
If yes, were there any problems with the drops? Please describe:		
Does he/she have difficulty seeing at night or in the dark?	Yes	No
Is he/she afraid of the dark or of shadows?	Yes	No
Does he/she have difficulty seeing the stars at night?	Yes	No
Does he/she have difficulty seeing sign language or gestures in dim light? (like at dusk)	Yes	No
Does he/she complain that bright lights hurt or bother him/her?	Yes	No
Does he/she need to wear sunglasses in order to see in bright sunlight?	Yes	No
When entering a new place or going from bright light to dim light or vice versa, does he/she ever stop suddenly, stand, and look around?	Yes	No
Does he/she ever confuse colors?	Yes	No
If yes, is the color problem with red & green? _____ yellow & blue?_____ dark colors like navy, black, or brown? _____		

BALANCE

At what age did he/she sit? ___ mo. crawl? _____ mo walk alone? _____ mo		
Did he/she have difficulty crawling or an abnormal crawl?	Yes	No
If yes, please describe how he/she crawled or moved about.		
Did he/she walk with the feet apart longer than most kids? If yes, until what age? _____ mo.	Yes	No
When learning to walk, most children sit down hard on their seats and catch themselves with their hands if they start to fall. Did your child do anything differently? If yes, describe what happened.	Yes	No
Did he/she wear out lots of shoes or trip because he/she "shuffled" instead of picking up the feet?	Yes	No
Do you think he/she is clumsier that other kids the same age? If yes, please describe.	Yes	No
When your child was 3-5 years old and you were going from the car to the house at night, did he/she: _____ stand or wait until someone came to take his/her hand? OR _____ walk to the house by him/herself?		
Does he/she have problems now walking on bumpy ground?	Yes	No
Does he/she often bump into objects like tables and chairs?	Yes	No
Does he/she often knock things over at mealtimes?	Yes	No
Does he/she stumble on stairs and curbs?	Yes	No
When he/she walks on a curb or 4-inch wide board, does he/she fall off? Don't know	Yes	No
Does he/she know how to ride a bicycle (two wheeler)? At what age did he/she learn how to ride? _____ yrs. How long did he/she need training wheels? _____	Yes	No
Does he/she know how to swim?	Yes	No
Does he/she have problems seeing stripes on the bottom of the pool? Don't know	Yes	No

OTHER HEALTH HISTORY

Did the mother have pregnancy problems? If yes, please explain..	Yes	No
Problems during delivery? If yes, please explain.	Yes	No
Newborn: Birth weight: Birth length: Head size:		
Was the child born prematurely? (too early) If yes, how many weeks early?	Yes	No
Did the baby have any problems like: _____ Yellow jaundice _____ Breathing problems _____ Meningitis _____ Other:		
How long did the baby stay in the hospital after birth? _____ days		
Has your child ever been in the hospital? If yes: DATE/AGE ILLNESS OR INJURY		
Does anything like soap, lotion, medicine, etc. make your child's skin red or swollen? If yes, what?	Yes	No
Has he/she ever been treated for seizures?	Yes	No
Has he/she ever been treated for fainting spells?	Yes	No
List any medications he/she regularly takes.		
Does he/she have any of the following? _____ White patch of hair _____ Kidney problems _____ White patches of skin _____ Goiter (large thyroid gland) or lump on neck _____ Arthritis _____ Diabetes _____ Bones break easily _____ Cleft lip (hare lip)/cleft palate (hole in roof of mouth) _____ Extra fingers or toes _____ Lip pits or dimples _____ Different colored eyes _____ Heart murmur or defect		

QUESTIONS ABOUT FAMILY

These questions are about your child's blood relatives, not relatives by marriage or adoption. The word "relation" in the tables below mean how that person is related to your child, not to you. When you answer these questions, please think about both the mother's and father's sides of the family, including your child's brothers, sisters, aunts, uncles, grandparents, great-grandparents, and cousins. Please print clearly.

Ethnic background (voluntary; check all that apply)

_____ European (what country?) _____
_____ Hispanic
_____ American Indian
_____ Other (what country?) _____
_____Asian (what country?) _____
_____ African American
_____ Pacific Islander

Does anyone in the family wear thick glasses, have problems seeing at night, or have any other eye trouble?

RELATION	KIND OF EYE PROBLEM	AGE WHEN PROBLEM STARTED

Does anyone in the family have a hearing problem (deaf, hard of hearing, sudden hearing loss, slow loss of hearing)?

RELATION	KIND OF HEARING PROBLEM	AGE WHEN PROBLEM STARTED

Does anyone in the family have neurological problems like seizures, dizziness, fainting spells, cerebral palsy, brain damage, or any other problem with the brain, spinal cord, or nerves?

RELATION	KIND OF NEUROLOGICAL PROBLEM	AGE WHEN PROBLEM STARTED

Who in the family does your child look like? _____

Are your child's mother and father related to each other? That is, are they cousins (even distant cousins)? Yes _____ No _____

Has your child or family ever seen a geneticist (a doctor who knows about inheritance or how problems are passed down in families)?

Yes _____ No _____

If yes, who was the geneticist and where (address)? _____

_____

Is there anything else we should know about your child?

_____

_____

_____

_____

Reprinted with permission from: Davenport, S. (unknown). *Usher screening project*. Bloomfield, MN: author.

KANSAS USHER SCREENING PROJECT

STUDENT QUESTIONNAIRE

Student:_____ Date of Birth: _____

Today's Date: _____

Note to Interviewer: Please take a minute to complete this questionnaire with your student or the student and the interpreter. This is to be completed for students from age 8 (or beginning in the third grade) through high school.

Students with Usher syndrome may often be the first to notice subtle changes associated with gradual vision loss. *Ask the student:*

- When you walk inside a dark movie theater or room, how long does it take you to adjust (e.g., a couple of seconds, one minute, or longer)?
- When you come inside from a bright, sunny day, is it hard to see things and people?
- Do you trip over things?
- Does sunlight hurt your eyes?
- Do you often spill liquids when pouring them into a cup or glass?
- Can you see the stars in the sky at night?
- If someone waves to you from the side, do you see them when you are not looking at them?
- Is it hard to find small objects when you drop them on the floor?
- Do you bump into people, chairs, or things?
- Do you have trouble riding a bicycle?
- Do you sometimes knock over a glass of water or other objects on the table at mealtimes?

Reprinted with permission from: North Dakota School for the Deaf. (1996). *Usher syndrome procedure.* Devils Lake, ND: Author.

KANSAS USHER SCREENING PROJECT

BEHAVIOR QUESTIONNAIRE

As a parent, you are in a unique position to detect symptoms of learning related vision problems in your child. These problems are not detectable by an eye chart test and can be overlooked during a quick screening. Please take a few minutes to help identify whether your child may require referral to an optometrist, ophthalmologist, or vision specialist. It should take less than 3 minutes to complete.

Name of child: _____

Birth date: _____ **Current date:** _____

Completed by: _____

If you have noticed any of the following behaviors, put an "X" in front of the statement. If a particular behavior does not apply, leave it blank.

Night Blindness

_____ can't see when coming in from bright sunlight
_____ trips over things when light changes or light is dim
_____ stays near light in a darkened room or at night
_____ positions self so light falls on the face of a speaker
_____ may express a desire to enter a room before it is darkened (e.g., movie theater)
_____ avoids conversations in darkened area
_____ may appear to stagger or lose balance after an oncoming car has passed at night
_____ has problems reading under some lights or in dimly lit areas

Visual Field

_____ stumbles on stairs and curbs
_____ bumps into people, tables, and chairs
_____ may bump or tip over objects placed to the side at mealtimes
_____ startles easily
_____ seems to hold objects in unusual positions when looking at them
_____ turns head while reading across a page
_____ uses fingers to mark place while reading
_____ can't find small objects that have been dropped
_____ fails to glance at another person's hand waving from the side
_____ is quiet or may edge to one side when in a large group
_____ frequently misses or fails to understand group instructions

Glare

_____ squints and shades eyes in bright lights or fluorescent lighting

_____ likes to wear sunglasses even in a building, but especially in bright sunlight

_____ may appear awkward when exiting a building (when faced with bright lights)

Contrast

_____ has difficulty reading light copies or ditto copies

_____ can't see stars at night

_____ often spills when pouring liquids

_____ eyes become watery and red when reading

Acuity

_____ holds book close to eyes or bends to read

_____ sits near blackboard

Balance

_____ late learning to walk (past 15 months)

_____ is considered clumsy

_____ loses balance easily in the dark

_____ can't ride a bicycle or required a long time to learn

Other

_____ is frequently last in completing group activities

_____ exhibits anxiety in new areas

_____ often last to enter a room

_____ may have repetitive behavior or routines at particular times

_____ may fail to participate fully in group activities associated with new situation in the dark

_____ frequently hesitates at the top or bottom of the stairs

_____ avoids walking or running in unfamiliar areas, especially in bright sunlight or dark area

_____ constantly appears to be visually scanning a group

_____ Does your child wear glasses or contacts?

If yes, _____ for reading only? _____ for distance only? _____ all the time?

Adapted from: Illinois Usher Screening Project. (unknown). *What you need to know about Usher syndrome*. Springfield, IL: Author.

Adapted from: Arizona Usher Screening Project. (1995). *Vision questionnaire*. Tucson: Arizona School for the Deaf and Blind.

KANSAS USHER SCREENING PROJECT

Student's Name: _____ **Age:** _____

Date: _____

CONE ADAPTATION

Procedure:

Scatter the squares from the Cone Adaptation Test in a jumbled heap on a dark table or carpet. The lights in the room should be dimmed to imitate twilight or dusk. The lighting needs to be even in the room. To see if the lights are dimmed to the correct level, ask if the student can see you signing If the student can see you sign, the room is too bright. Ask the student to pick up the white squares. Note how long the student takes to complete the task. Then ask the student to sort the red squares from the blue squares. Document the start time, stop time, and total time it took to complete the task. Document the number of squares correctly sorted. Conduct this test two times for each student.

Results:

Trial #1

_____ Time started

_____ Timed stopped

_____ Total time

_____ _____ _____ Number correct

(white) (blue) (red)

Trial #2

_____ Time started

_____ Timed stopped

_____ Total time

_____ _____ _____ Number correct

(white) (blue) (red)

Adapted from: North Dakota School for the Deaf (1996). *Dark adaptation test.* Devils Lake, ND: Author.

KANSAS USHER SCREENING PROJECT

Student's Name: _____ **Age:** _____

Date: _____

FIELD SCREENING

Procedure: Conduct either the Wiggling Fingers Test or the Two Fingers Test.

Wiggling Fingers Test
This test should be conducted with both of the student's eyes open. You and the student should be seated facing each other at approximately one arm's length. Tell the student to look at your nose and tell you when he or she sees your fingers. Extend both of your arms to the side of your body. Bend both of your wrists and begin wiggling your fingers while moving your arms closer to the student. Document at which point the student can first see your wiggling fingers. Continue bringing your hands in closer toward your body to detect for ring scotomas. Conduct the test for both horizontal and vertical planes.

Two Fingers Test
This test should be conducted with both of the student's eyes open. You and the student should be seated facing each other at approximately one arm's length. Tell the student to look at your nose. Hold one or two fingers to the side of your own visual field. Ask the student whether one or two fingers are present. Document at which point the student can see the fingers. Continue bringing your hands in closer toward your body to detect for ring scotomas. Conduct the test for both horizontal and vertical planes.

Results: Color in the areas the student was able to see. Use a dark color (not a highlighter). Any section of the semicircle not colored in means that the student failed the screening.

Horizontal

_____ both eyes

CHILD

Results: Color in the areas the student was able to see. Use a dark color (not a highlighter). Any section of the semicircle not colored in means that the student failed the screening.

Vertical

_____ both eyes

CHILD

Adapted from: Davenport, S. (1996). *Vision screening project.* Bloomington, MN. Author.
Adapted from: Alaska Center for Blind and Deaf Adults & Alaska Services for Children and Youth with Sensory Impairments. (1994). *Usher screening forms.* Anchorage: Author.

KANSAS USHER SCREENING PROJECT

Student's Name: _____ **Age:** _____

Date: _____

BALANCE

Procedure: Conduct both of these tests.

Feet Together
Have the student stand with feet together, arms stretched out to the side of the body, and eyes closed. Stand behind the student while gently pushing on either side of the student's torso. (Note: the student may lose balance quickly so be ready to support him or her.) Document whether the student was able to maintain his or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained	Balance not maintained	
_____	_____	eyes closed with feet together
_____	_____	eyes open with feet together

One Foot in Front of the Other
Have the student stand with one foot in front of the other, arms stretched out to the side of the body, and eyes closed. Stand behind the student while gently pushing on either side of the student's torso. (Note: the student may lose balance quickly so be ready to support him or her.) Document whether the student was able to maintain his or her balance. Conduct the same procedure again, but this time the student's eyes need to be open. Document the student's reaction.

Balance maintained	Balance not maintained	
_____	_____	eyes closed with one foot in front of the other
_____	_____	eyes open with one foot in front of the other

Adapted from: North Dakota School for the Deaf (1996). *Balance*. Devils Lake, ND: Author.

KANSAS USHER SCREENING PROJECT

USHER SCREENING DATA SUMMARY

Student _____ Birth date _____ Name of district or school _____

Person completing data summary form _____ Date completed _____

	COMPLETED?		WHY NOT?
Family Questionnaire	YES	NO	
Student Questionnaire	YES	NO	
Behavior Questionnaire from:			
parents	YES	NO	
teacher	YES	NO	
dorm teacher	YES	NO	
Behaviors	YES	NO	
Night blindness	YES	NO	
Visual field	YES	NO	
Glare	YES	NO	
Contrast	YES	NO	
Acuity	YES	NO	
Balance	YES	NO	
Other	YES	NO	

	COMPLETED?		APPEARS NORMAL?	
Cone Adaptation	YES	NO	YES	NO
Field Screening	YES	NO	YES	NO
Balance Screening	YES	NO	YES	NO

	RECOMMEND SENDING?		TO WHOM?
Referral letter	YES	NO	

	COMPLETED?		TO WHOM?
Followup	YES	NO	

Comments:

PROYECTO DE DETECCION DEL SINDROME DE USHER DEL ESTADO DE KANSAS

CUESTIONARIO DEL ESTUDIANTE

Estudiante: _____ **Fecha de Nacimiento:** _____

Fecha de hoy: _____

Nota para el Entrevistador: Por favor tome un minuto para completar este cuestionario con su estudiante o él y su intérprete. Esto es para completar por estudiantes desde la edad de 8 años (o que estén empezando el tercer grado) hasta escuela superior.

Los estudiantes con Síndrome de Usher frecuentemente pueden ser los primeros en notar cambios leves asociados con la pérdida gradual de visión. Preguntar al estudiante:

- ¿Cuando entra al teatro o cine o a un cuarto oscuro, cuánto tiempo tarda en ajustarse (ej., unos segundos, un minuto, o más)?
- ¿Cuando Ud. entra despues de estar fuera en un día soleado y brillante, le es dificil ver cosas y gente?
- ¿Se tropieza sobre cosas?
- ¿La luz del sol lastima sus ojos?
- ¿Derrama los líquidos frecuentemente cuando los vierte o sirve en una taza o vaso?
- ¿ Puede ver las estrellas en el cielo de noche?
- ¿Si alguien le hace señas por un lado, los puéde ver aunque no los esté mirando directamente?
- ¿Se le dificulta encontrar objetos pequeños cuando se le caen al piso?
- ¿Se tropieza con gente, sillas, o cosas?
- ¿Se le dificulta andar en bicicleta?
- ¿Algunas veces tira sobre la mesa un vaso de agua u otros objetos durante las comidas?

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PROYECTO DE DETECCION DEL SINDROME DE USHER DEL ESTADO DE KANSAS

CUESTIONARIO DE COMPORTAMIENTO

Como padre, Ud. está en posición única de detectar síntomas y notar los problemas relacionados con la vista en su niño. Estos problemas no se detectan por un examen común de ojos y pueden no detectarse durante una selección rápida. Por favor tomar unos minutos para ayudar a identificar si su niño puede necesitar referencia a un optometrista, oftalmólogo, o especialista de vista, debe tomar menos de 3 minutos para completar.

Nombre de niño: _____

Fecha de nacimiento: _____ Fecha actual: _____

Completado por: _____

Si Ud. ha notado cualquiera de los comportamientos siguientes, ponga una "X" en frente de la declaración. Si un comportamiento particular no aplica, déjelo en blanco.

Ceguera de noche

_____ No poder ver cuando entra después de estar en un sol brillante
_____ Tropieza sobre cosas cuando la luz brillante cambia o es opaca
_____ Permanece cerca de la luz en una sala oscura o de noche
_____ Se ubica de tal manera que la luz alumbre la cara de la persona que esta hablando
_____ Expresa su deseo de entrar en una sala antes de que apagen las luces (ej. teatro de cine)
_____ Evita conversaciones en áreas oscuras
_____ Parece tambalear o perder equilibrio después de que un automóvil dirigido hacia el pasa en la noche
_____ Tiene problemas al leer en áreas bajo luces o ligeramente iluminadas

Campo Visual

_____ Tropieza sobre escaleras y banquetas
_____ Tropieza con gente, mesas, y sillas
_____ Tropieza o tira objetos puestos a su lado durante las comidas
_____ Se sorprende fácilmente
_____ Parece coger los objetos en posiciones inusitadas cuando los vé
_____ Mueve la cabeza cuando esta leyendo una página
_____ Usa sus dedos para marcar el lugar mientras está leyendo
_____ No puede encontrar objetos pequeños que se han caido
_____ No vé cuando otra persona le hace señas con la mano por un lado
_____ Es callado o se hace a un lado cuando está con un grupo grande
_____ Frecuentemente no se dá cuenta o no puede comprender instrucciones en grupo

Reflejo

_____ casi cierra los ojos o parpadea en luz brillante o fluorescente
_____ le gusta usar lentes oscuros aún adentro, pero especialmente en sol brillante
_____ puede parecer torpe al salir de un edificio (cuando encara con luz brillante)

Contraste

_____ tiene dificultad al leer copias no muy claras o copias ditto
_____ no puede ver las estrellas de noche
_____ frecuentemente cuando vierte líquidos los derrama
_____ los ojos le lloran y se le ponen rojos cuando lee

Acuidad

_____ pone el libro cerca de sus ojos o se agacha para leer
_____ se sienta cerca del pizarrón

Equilibrio

_____ aprendió tarde a caminar (pasando 15 meses)
_____ se considera torpe
_____ pierde fácilmente el equilibrío en la oscuridad
_____ no puede andar en bicicleta o requirió un tiempo largo para aprender

Otros

_____ frecuentemente es el último en completar actividades del grupo
_____ expresa ansiedad en áreas nuevas
_____ frecuentemente es el último en entrar en una sala
_____ puede tener comportamiento o rutinas repetidos en tiempos particulares
_____ no puede participar totalmente en actividades de grupo asociadas con una situación nueva en la oscuridad
_____ frecuentemente duda al bajar o subir las escaleras
_____ evita caminar o correr en áreas no conocidas, especialmente áreas brillante soleiadas u obscuras
_____ constantemente aparece repasando un grupo visualmente

_____ ¿ Usa su niño anteojos o lentes de contacto?
Si sí,___ para leer únicamente? ___ para la distancia únicamente? ___ todo el tiempo?

Adapted from: Illinois Usher Screening Project. (unknown). What you need to know about Usher Syndrome. Springfield, IL: Author.

Adapted from: Arizona Usher Screening Project. (1995). Vision questionnaire. Tucson: Arizona School for the Deaf and Blind.

PROYECTO DE DETECCION DEL SINDROME DE USHER DEL ESTADO DE KANSAS

Proyecto de Detección del Sindrome Usher para Niños Sordos o con Perdida de Oír

CUESTIONARIO DE LA FAMILIA

Nombre del niño	Fecha de Nacimiento	Fecha de hoy
Dirección Teléfono		
Nombre de la persona que está llenando esta forma:	Relación con el estudiante:	

PREGUNTAS ACERCA DEL NINO

AUDIENCIA

¿Que edad tenía su niño cuando quedó sordo? _____ años		
¿Por qué el o ella quedó sordo?		
¿El o ella ha usado aparatos auditivos?	Sí	No
¿Si sí, los aparatos auditivos ayudaron?	Sí	No
¿La audiencia de su niño ha sido mejor antes de ahora?	Sí	No

VISTA

¿Ha usado lentes alguna vez su niño?	Sí	No
¿Si sí, que edad tenia su niño cuando empezó a usarlos? _____años		
¿El o ella tiene problemas al ver objetos de lejos, pero vé los de cerca (miope)?	Sí	No
¿Su niño tiene problema ver de cerca, pero vé bien de lejos (hipermétrope)?	Sí	No
¿Alguna vez a su niño le han puesto gotas para los ojos para dilatarlos?	Sí	No
¿Si sí, había cualquier problema como resultado de las gotas? Por favor describir:		
¿El o ella tiene dificultad de ver de noche o en la oscuridad?	Sí	No
¿El o ella tiene miedo de la oscuridad o de sombras?	Sí	No
¿El o ella tiene dificultad de ver las estrellas de noche?	Sí	No
¿El o ella tiene dificultad de ver gestos o lenguaje de señas en la luz opaca? (como en el anochecer)	Sí	No
¿El o ella se queja que la luz brillante le lastima o le molesta?	Sí	No
¿El o ella necesita usar lentes oscuros para poder ver en la luz de un sol brillante?	Sí	No
¿Cuando entra en un lugar nuevo o vá de luz brillante a opaca o viceversa, él o ella para repentinamente, y mira a su alrededor?	Sí	No
¿El o ella confunde colores?	Sí	No
¿Si sí, es el problema de color con el rojo & verde?___ amarillo & azul?___ colores obscuros como azul marino, negro, o marrón?___		

EQUILIBRIO

¿A qué edad se sentó su niño? ____ meses gateó? ____ meses camino solo? ____ meses		
¿El o ella tuvo dificultad al gatear o un gateo anormal?	Sí	No
Si sí, por favor describir como él o ella gateó o se movió.		
¿El o ella caminó con los pies separados por más tiempo que la mayoría de niños? ¿Si sí, hasta qué edad? ____ meses.	Sí	No
La mayoría de los niños cuando aprenden a caminar se dan sentones duros pero se detienen con las manos a caer. ¿Hizo algo diferente su niño? Si sí, describir qué sucedió.	Sí	No
El o ella acaba pronto los zapatos o se tropieza porque él o ella "arrastra" los pies en vez de alsarlos?	Sí	No
¿Ud. piensa que él o ella es más torpe que otros niños de la misma edad? Si sí, por favor describir.	Sí	No
¿Cuando su niño tenía 3-5 años de edad y Ud. fué de su automóvil a la casa de noche él o ella: _____ esperó hasta que alguien vino para tomar su mano? O _____ caminó a la casa por si mismo?		
¿Ahora él o ella tiene problemas al caminar sobre terreno irregular?	Sí	No
¿El o ella frecuentemente tropieza con objetos tales como mesas y sillas?	Sí	No
¿El o ella frecuentemente tira cosas sobre la mesa a la hora de comer?	Sí	No
¿El o ella tropieza sobre escaleras y banquetas?	Sí	No
¿Cuando él o ella camina sobre una banqueta o una tabla de 4 pulgadas, él o ella se cae afuera? ____ no sabe	Sí	No
¿El o ella sabe como andar en bicicleta (de dos ruedas)? ¿A qué edad él o ella aprendió? _____años ¿Cuánto tiempo él o ella necesitó ruedas de entrenar? _____ ¿El o ella sabe como nadar?	Sí Sí	No No
¿El o ella tiene dificultad de ver las rayas sobre el fondo de la piscina? ____ no sabe	Sí	No

OTRA HISTORIA DE SALUD

¿La madre tuvo problemas durante el embarazo? Si sí, por favor explicar.	Sí	No
¿Problemas durante el nacimiento? Si sí, por favor explicar.	Sí	No
Recien nacido: Peso al nacer: Tamaño al nacer: Medida de la cabeza:		
¿Nació el niño prematuramente? (demasiado temprano) ¿Si sí, cuántos semanas antes?	Sí	No
¿El bebé tuvo algun problema como: _____ Ictericia amarilla _____ Problemas respiratorios _____ Meningitis _____ Otro:		
¿Cuánto tiempo permaneció el bebé en el hospital después de su nacimiento? _____ días		
¿Ha estado su niño en el hospital? Si sí: FECHA/ EDAD DANO O ENFERMEDAD		
¿Alguna cosa como jabón, loción, medicina, etc. hace que la piel de su niño se ponga roja o hinchada? ¿Si sí, qué es?	Sí	No
¿El o ella ha sido tratado de ataques alguna vez?	Sí	No
¿El o ella he sido tratado por desmayos?	Sí	No
Notar cualquier medicacion que él o ella toma regularmente.		
¿El o ella tiene cualquiera de lo siguiente? _____ Mechón blanco de pelo _____ Manchas Blancas en la piel _____ Artritis _____ Huesos que rompen fácilmente _____ Dedos extras de la mano o del pie _____ Ojos de color diferente _____ Problemas de riñón _____ Bocio (glándula tiroidea grande) o protusíon en el cuello _____ Diabetes _____ Labio partido (labio de liebre)/ paladar partido (hoyo en el paladar) _____ el Labio endido u hoyuelos _____ Murmullo o falla en el corazón		

PREGUNTAS ACERCA DE LA FAMILIA

Estas preguntas son sobre parientes cercanos de su niño, no parientes por casamiento o adopción. La palabra "la relación" abajo en los renglones significa como esa persona se relaciona a su niño, no a Ud. Cuando Ud. conteste estas preguntas, por favor piense en ambos la madre y el padre de la familia, incluyendo hermanos y hermanas de su niño, tías, tíos, abuelos, bisabuelos, y primos.

Por favor escriba claramente.

Antecedentes étnicos (voluntariamente; checar todo lo que aplica)

_____ Europeo (qué país?) _____
_____ Hispano (qué país?) _____
_____ Indio Americano
_____ Otro (qué país?) _____

___ Asiático (qué país?)___ _____
___ Africano Americano
___ Isleño del Pacífico (qué país?)_____ ____

¿Alguien en la familia usa anteojos gruesos, tiene problemas de ver de noche, o tiene algún otro problema de vista?

RELACION	TIPO DE PROBLEMA DE VISTA	EDAD CUANDO EMPEZO EL PROBLEMA

¿Alguien en la familia tene problema de oido (sordo, un poco sordo, pérdida de repente o pérdida lenta de oido)?

RELACION	TIPO DE PROBLEMA DEL OIDO	EDAD CUANDO EMPEZO EL PROBLEMA

¿Alguien en la familia tene problemas neurológicos como, ataques, mareos, desmayos, paralisis cerebral, daño de cerebro o algún otro problema de cerebro, cordón espinal, o nervios?

RELACION	TIPO DE PROBLEMA NEUROLOGICO	EDAD CUANDO EMPEZO EL PROBLEMA

¿A quien en la familia se parece su niño?

¿Son parientes los padres de su niño? ¿Esto es, son primos (o distantes primos)? Sí___ No___

¿Alguna vez Ud o su niño han visto un genetista (un doctor que sabe sobre herencia o como problemas son pasados entre familiares)? Sí___ No___

¿Si sí, quien es el genetista y cual es su dirección?

¿Hay alguna otra cosa que nosotros deberíamos saber sobre su niño?

_____
_____
_____
_____

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